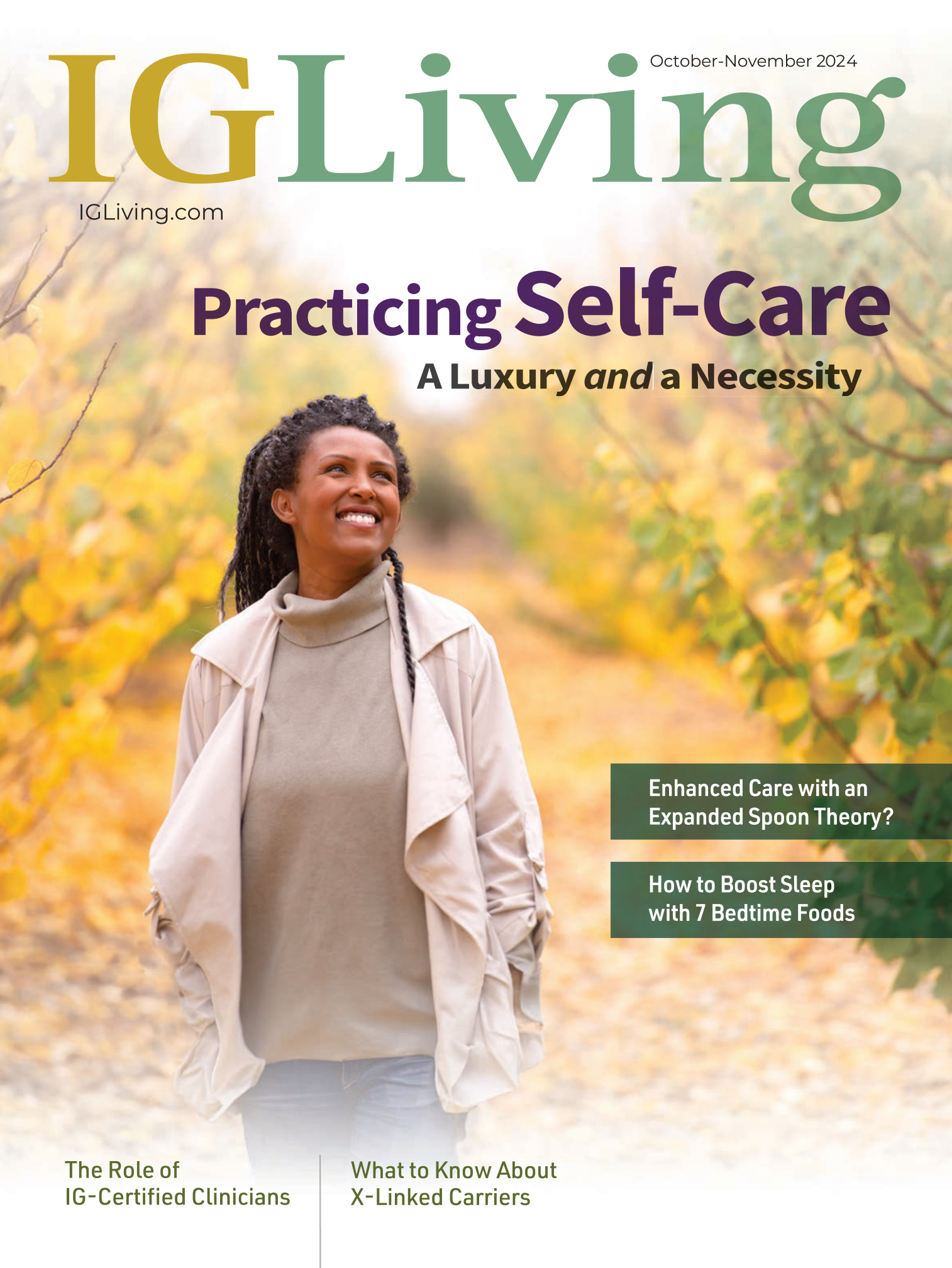
October-November 2024

# IGLiving

IGLiving.com

# Practicing Self-Care

## A Luxury *and* a Necessity



Enhanced Care with an Expanded Spoon Theory?

How to Boost Sleep with 7 Bedtime Foods

The Role of IG-Certified Clinicians

What to Know About X-Linked Carriers

FOR PATIENTS WITH PRIMARY HUMORAL IMMUNODEFICIENCY (PI)

# IT'S WHAT'S INSIDE THAT COUNTS

ASCENIV™

IMMUNE GLOBULIN INTRAVENOUS (HUMAN) – slra 10% LIQUID

**DESIGNED TO DELIVER**

**Talk to your doctor about whether ASCENIV™ is right for you** asceniv.com

Important Safety Information for ASCENIV™

WARNING: RISK OF BLOOD CLOTS (THROMBOSIS), POOR KIDNEY FUNCTION, AND INABILITY TO FILTER WASTE FROM KIDNEYS. BLOOD CLOTS MAY OCCUR WITH INTRAVENOUS IMMUNE GLOBULIN PRODUCTS, INCLUDING ASCENIV.

Before taking ASCENIV, talk to your doctor if you:

- Are of advanced age
- Are unusually sedentary (long periods of sitting down or inactive)
- Are taking estrogen-containing medicines (birth control pills, hormone replacement therapy)
- Have a permanent intravenous (IV) catheter
- Have hyperviscosity of the blood (diseases such as multiple myeloma or other causes of elevated proteins in the blood)
- Have cardiovascular (heart) problems or previous history of stroke

Thrombosis may occur even if you do not have any risk factors.

Serious kidney problems and death can also happen in certain patients who receive such products.

If you are at high risk of thrombosis or kidney problems, your doctor should adjust the dose of ASCENIV and will monitor you for signs and symptoms of thrombosis and viscosity, as well as kidney function.

**What is ASCENIV (immune globulin intravenous, human)?**
ASCENIV (immune globulin intravenous, human) is a prescription medicine to help adults and adolescents (12 to 17 years old) with primary immunodeficiency fight and prevent infections.

ASCENIV is for intravenous administration only. ASCENIV is made from healthy human blood/plasma.

**Who should not use ASCENIV?**
ASCENIV should not be used if you had a severe allergic reaction to human immune globulin or if you have been told by a doctor that you are immunoglobulin A (IgA)-deficient and have developed antibodies to IgA and hypersensitivity after exposure to a previous plasma product.

**What are possible warnings and precautions with taking ASCENIV?**
**Hypersensitivity.** Severe allergic reactions may occur with immune globulin products, including ASCENIV. If you have a severe allergic reaction, stop the infusion immediately and get medical attention. ASCENIV contains IgA. If you have known antibodies to IgA, you may have a greater risk of developing potentially severe allergic reactions.

If you take ASCENIV or a similar immune globulin product, you could experience a serious and life-threatening blood clot (thromboembolism). This may include pain and/or swelling of an arm or leg with warmth over the affected area, discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness, or weakness on one side of the body. If you are at risk, your doctor may decide to adjust the dose of ASCENIV. Your doctor will monitor you for any signs or symptoms of blood clots or poor blood flow in your arteries.

**Always tell your doctor immediately if your medical history is similar to what is described here, and especially if you experience any of these symptoms while taking ASCENIV.**

**Kidney problems or failure.** Kidney problems, kidney failure, and death may occur with use of human immune globulin products, especially those containing sucrose (sugar). ASCENIV does not contain sucrose.

If you have kidney disease or diseases with kidney involvement, your doctor should perform a blood test to assess your hydration level and kidney function before beginning immune globulin treatment and at appropriate intervals thereafter. If your doctor determines that kidney function is worsening, they may discontinue treatment. If your doctor determines you to be at risk, they may start your dose of ASCENIV at a safe level.

**People taking human immune globulin products, including ASCENIV, may experience hyperproteinemia (high levels of protein in the blood), hyponatremia (low levels of sodium in the blood), and hyperviscosity (poor blood flow). Your doctor may perform certain blood tests and monitor you to minimize any of the above risks.**

**Aseptic meningitis syndrome (AMS).** Aseptic meningitis is a non-infectious inflammation of the membranes that cover the brain. It causes a severe headache, which may occur with human immune globulin treatment, including ASCENIV. AMS usually happens within a few hours to 2 days after treatment. AMS is more commonly associated with higher doses of treatment and/or after rapid infusion. Your doctor may perform a neurological exam, including spinal tap (sampling fluid which surrounds the spinal cord) to evaluate your condition and to rule out other causes of meningitis.

**Hemolysis.** Hemolysis refers to the destruction of red blood cells. Immune globulin products, including ASCENIV, may contain certain antibodies that can result in the rupturing of red blood cells. Your doctor should monitor you for signs and symptoms of hemolysis, which may include additional confirmation tests.

Taking intravenous human immune globulin products may cause a build up of fluid in the lungs (pulmonary edema) that is unrelated to heart problems. Your doctor should monitor you for lung-related side effects and may conduct appropriate tests that can detect the presence of certain white blood cells (anti-neutrophils) in the drug or your blood. If needed, your doctor may decide to use oxygen or other respiratory methods to help your breathing.

**Transmissible infectious agents.** Because ASCENIV is made from human blood, it may carry a risk of transmitting infectious agents such as viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent. Your doctor will report to the manufacturer any cases of suspected infections spread by the product.

**Interference with lab tests.** Because ASCENIV contains a variety of antibodies that are infused into your body, blood tests to determine antibody levels may provide misleading interpretations. Be sure to always tell your doctor, nurse, or lab technician of any medicines you are taking and that you are using ASCENIV.

**Interactions with medicines.** ASCENIV can make vaccines (like measles, mumps, rubella, and chicken pox vaccines) less effective in your body. Before you get any vaccines, tell your healthcare provider that you take ASCENIV.

**What are other possible side effects of ASCENIV?**
In clinical studies of ASCENIV, some patients experienced the following:

- Headache
- Sinus inflammation (sinusitis)
- Diarrhea
- Intestinal lining inflammation caused by virus (gastroenteritis)
- Common cold (nasopharyngitis)
- Upper respiratory tract infection
- Bronchitis
- Nausea

**These are not all the possible side effects of ASCENIV. Talk to your healthcare provider about any side effect that bothers you or that does not go away.**

**You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch or call 1-800-FDA-1088.**

**For additional safety information about ASCENIV, please see full Prescribing Information at www.asceniv.com**

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# Features



# Up Front



# Departments



# Columns



# Sources





**About IG Living**
IG Living magazine brings together patients, advocates and caregivers in the immune globulin (IG) community.


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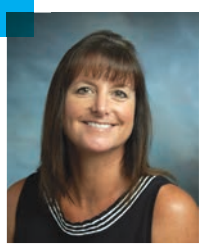
# Prioritizing and Practicing Self-Care



**PRIORITIZING SELF**-care can revitalize your energy and ensure you're ready to handle whatever comes your way in a safe and healthy way. But, prioritizing and practicing self-care isn't always easy. Sometimes, self-care is misunderstood for adding tasks to your already overflowing plate. But, the truth is, self-care is more about doing less than more and being less concerned about disappointing others and more concerned with loving *and saving* yourself.

This is why we titled our cover feature "Why Self-Care Is Anything But Selfish" (p.24) because while many chronically ill patients *feel* self-care is somehow selfish, it *isn't*. Rather, it's a critical aspect of maintaining your physical and mental health. As we explain in the article, a well-balanced self-care routine involves seven basic pillars — mental, emotional, physical, environmental, spiritual, recreational and social — adapted to each individual's needs. Yes, that can mean adding activities such as going to a concert with others to socialize, but it also means reducing stress levels by focusing on what is most physically and mentally restorative for you such as journaling your feelings and doing what matters most to you. The keys are to reduce risk factors, increase protective factors and practice positivity.

Practicing self-care is a fundamental aspect of the spoon theory. Coined in a 2003 essay by American writer Christine Miserandino, the spoon theory is a metaphor using spoons to describe the amount of physical or mental energy a person has available for daily activities and tasks, and how these spoons can become limited for those with chronic illnesses versus healthy individuals. In our article "The Spoon Theory, Reimagined?" (p.28), we take a look at how the original concept of the spoon theory might be expanded beyond balancing tasks and empowering patients, to exploring innovative approaches to the concept such as redefining energy and resources, incorporating accessibility and inclusivity, providing tools for advocacy, leveraging technology and more.

In the end, self-care is all about taking care of yourself and giving your body and mind what it needs. And, that includes a good night's sleep! So, registered dietitian Emily Cooper shares her tips for adding specific foods to your bedtime routine in her article "7 Foods for a Better Night's Sleep" (p.36). She also provides a list of foods to avoid later in the day because they could potentially hinder your sleep.

As always, we hope you enjoy these articles, as well as the many more educational and insightful topics presented in this issue of *IG Living*.

Ronale Tucker Rhodes

Ronale Tucker Rhodes, MS

IGLiving

Our mission is to support the IG community through education, communication and advocacy

A community service from FFF Enterprises, Inc.

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# Resources and Assistance Programs for Patients Undergoing IG Therapy

By Abbie Cornett, MBA


**MANY PATIENTS** frequently ask me where they can find resources and assistance programs that can support them and their families in navigating their immune globulin (IG) treatment journey.

One of the primary organizations offering support and information for rare diseases, including conditions requiring IG therapy, is the National Organization for Rare Disorders (NORD). NORD provides patients with the necessary resources, support and advocacy to manage their conditions effectively, as well as patient assistance programs that offer additional resources and support.

The Immune Deficiency Foundation (IDF) is another crucial resource, particularly for patients treated with IG therapy. While IDF primarily focuses on primary immune deficiencies, it also provides insurance advocacy assistance to help patients navigate the complexities of their treatment and coverage.

Another significant organization is the GBS|CIDP Foundation International, which supports patients with Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy and other variants. This foundation provides resources and patient support, ensuring those affected by these conditions receive the necessary information and assistance.

Managing the costs associated with IG therapy can be a significant burden. Fortunately, several organizations offer co-payment assistance programs, including the Patient Access Network Foundation, which provides financial assistance to underinsured patients, helping cover out-of-pocket costs associated with IG therapy, and the HealthWell Foundation, which offers co-payment assistance for patients, including those requiring IG therapy.

Many pharmaceutical companies that manufacture IG products have established patient assistance programs to help cover the cost of IG therapy for eligible patients. Companies such as CSL Behring, Grifols and Takeda offer programs tailored to the specific needs of patients, ensuring they receive the necessary treatments without undue financial hardship. Another valuable resource is Accessia Health, formerly Patient Services Inc., which provides premium and co-payment assistance for patients, including those on IG therapy.

Navigating insurance policies and coverage can be a daunting task for many patients and their families. One of the best strategies is for patients to regularly review their insurance policy to understand coverage and any changes that may occur. Engaging with patient advocates or case managers can also be beneficial to help navigate insurance complexities and provide valuable guidance.

Maintaining detailed records of the diagnosis, treatment history and the necessity of IG therapy can be essential in appealing insurance denials. These records provide the necessary evidence to support the need for treatment, making it easier to challenge insurance company decisions.

When insurance companies deny coverage, seeking legal assistance may be necessary. Organizations such as the Patient Advocate Foundation can provide guidance and support and help patients understand their rights and explore options for appealing insurance denials.

Participating in support groups, both online and in-person, can ease anxiety and provide a source of comfort and understanding. Connecting with others who are going through similar experiences helps patients feel less isolated and more supported in their journey. Speaking with a mental health professional, who can provide strategies to manage anxiety and fear related to the medical condition and treatment, can also be beneficial. By addressing emotional challenges, patients can better cope with their condition and maintain a more positive outlook.

By leveraging the many available resources, patients and their families can navigate the complexities of their condition more effectively, ensuring access to necessary treatments and reducing the associated financial and emotional burdens. IG

**ABBIE CORNETT,** MBA, is the patient advocate for *IG Living* magazine. She can be reached at patient advocate@igliving.com or (800) 843-7477 x1366.


## What Lessons Have You Learned Navigating Life with Chronic Illness?

*That it's never easy, and every new disease and diagnosis I get is just added to the pile. I started talking to a therapist on the phone for Telehealth. I did it just about every week all last year, and then she was out for surgery and then she had another surgery in March, so now I'm waiting to get assigned to somebody else. It's helped a lot with my stress and anxiety with my myriad of health issues and my family issues and all the drama. It's free because I have Medicare and it's through my Medicare Advantage plan. Years ago, I did cognitive behavioral therapy with a psychologist and it was a book on how to manage chronic pain before it manages you, so this is kind of the same thing with a therapist now — to learn to say no to everything. And then she gave me the acronym "Jade." It's when somebody invites you to something; there's no reason to justify, acknowledge, describe or explain why you said no to a friend, to a family event, etc.*

*Focus on the good things in life. Count your blessings. Find happy things to focus on each day. Don't let chronic illness make you distraught. My faith in God gives me strength to do all things!*

## Do You Have a Hobby?

*Many! Good distractions from all of this health stuff!*

*Gym workouts, reading, walking, landscaping.*

*Scrapbooking*

*Yes, I'm a writer.*

*I crochet, make jewelry, do some metalworking and paint. As a disabled person, my hobbies are an important part of keeping me sane. They also help me to focus on something other than my illnesses.*


## When Does Toxic Positivity Not Work?

*Toxic positivity is when reality is completely negated. It's blindly speaking positively and ignoring what's actually happening. But toxic positivity doesn't miraculously make everything better. In fact, it makes everything worse because it dismisses reality and invalidates people's individual experience.*

*I try to find the best in things, but completely burying any emotions that aren't considered positive doesn't help you in the long run. At this point, I'd far rather feel whatever I happen to be feeling at the time, instead of pasting a completely fabricated smile on my face 24/7.*

**Join the conversation!** Connect with other immune globulin patients through IG Living's Facebook page at www.facebook.com/IGLivingMagazine. Each day, we post interesting articles and facts, as well as thought-provoking questions you can weigh in on. These are some snapshots of what's being discussed.

## Can Ablation of Spine Cause a Flare-Up of CIDP?

I've been dealing with pain from a thoracic spine injury and had kyphoplasty (a procedure to treat compression fractures) this past June; however, it was unsuccessful. I have been in remission for 10 years with chronic inflammatory demyelinating polyneuropathy (CIDP). It has been recommended that I receive ablation of spine to burn four nerves if my diagnostic is positive. But, could this procedure flare up my CIDP?

**Abbie:** I spoke with Terry O. Harville, MD, PhD, medical director of the Special Immunology Laboratory at the University of Arkansas for Medical Sciences, and he said your concern is valid. He explained that the exact cause of CIDP is not fully understood, but it likely involves nerve injury, inflammation and autoimmunity. Unfortunately, nerve injury during procedures can precipitate another event.

He suggested several alternatives that might be beneficial: 1) local corticosteroid injection, which could reduce local inflammation and provide relief; 2) local nerve block, an anesthetic agent that can offer long-term relief; 3) acupuncture and TENS, therapies that can provide relief for some individuals; and 4) low-dose naltrexone and celecoxib, which are being successfully used in some situations to reduce central nervous system inflammation and body pain.

If these alternatives are not effective and the spinal ablation procedure is necessary, he recommends a pretreatment plan to potentially prevent a CIDP flare, including 1 gram of intravenous methylprednisolone three days before the procedure and 1 gram of intravenous methylprednisolone immediately before the procedure. This could dampen the inflammatory and immune responses to the acute injury.

Dr. Harville does recommend you discuss and individualize this plan with your physicians.

## Are There Resources if IVIG Is Denied for Autoimmune Small Fiber Polyneuropathy?

I am a physician and the mother of an adult daughter who has been diagnosed with autoimmune small fiber polyneuropathy. She has been receiving monthly intravenous immune globulin (IVIG) infusions for the past several years and has done very well on this regimen. Before being administered IVIG, she was extremely symptomatic and her quality of life was not good.

As you might imagine, she is fearful that her insurance company will at some point refuse to cover the cost of her infusions. Any information you can provide regarding resources available to her should this occur will be greatly appreciated and may help alleviate her anxiety.

**Abbie:** There are a number of resources and assistance programs for patients with autoimmune small fiber polyneuropathy, particularly concerning IVIG therapy:

Information and support: 1) National Organization for Rare Disorders (NORD): rarediseases.org; 2) GBS|CIDP Foundation International: www.gbs-cidp.org; 3) Immune Deficiency Foundation: primaryimmune.org.

Co-payment assistance programs: 1) Patient Access Network (PAN) Foundation: www.panfoundation.org; 2) HealthWell Foundation: www.healthwellfoundation.org.

Patient assistance programs: 1) Many pharmaceutical companies provide assistance programs, including CSL Behring, Grifols and Takeda, to help cover the cost of IVIG for eligible patients; 2) Patient Services Incorporated (PSI) provides premium and co-payment assistance for patients with chronic illnesses, including those on IVIG therapy: www.patientservicesinc.org.

**» Have a question?** Email us at editor@IGLiving.com.
Your information will remain confidential unless permission is given.

ABBIE CORNETT, MBA, is the patient advocate for *IG Living* magazine. She can be reached at patient advocate@igliving.com or (800) 843-7477 x1366.

# Balancing Gratitude and Grief

By Mairead McConnell, PhD

**AS WE SETTLE** into autumn and prepare for the holidays, the term "gratitude" is getting a lot of attention. Gratitude is the practice or attitude of being thankful, of being appreciative. Not only is gratitude lauded by society as an important virtue, but some studies have found gratitude to be associated with greater well-being and physical health.[1] But is gratitude really that simple? Can you just appreciate your way into better health, or a better life?

The expectation of gratitude can feel burdensome when we consider the pain and suffering that often characterizes our lives and world. When life feels especially difficult, when a loved one dies far too soon, when illness strikes, when wars rage around the world or hit too close to home — gratitude may, or may not, come so readily. As you navigate this season, consider these ways to hold gratitude in balance with whatever else you are feeling.

*As you navigate this season, consider these ways to hold gratitude in balance with whatever else you are feeling.*

## Don't Force It

"Just be grateful!" "Look on the bright side." "It could be worse." We've all seen the bumper stickers or heard these comments from a well-meaning friend. And yet, forcing gratitude or positivity is another way we dismiss and invalidate one another and even ourselves. More important than forcing gratitude is acceptance — acknowledging that however we feel is OK and makes sense. Sometimes — often — it is perfectly valid to want more or to want to heal our emotional wounds, feel more physically well or to live in a more just world. You are not ungrateful for wanting more. You're human.

## The Myth of Ungrateful

Almost as pervasive as the word gratitude is the label "ungrateful." This term is often reserved for children who fail to display an expected level of appreciation, but it can also be used as an insult for adults who do the same. I urge you to consider that there is no such thing as ungrateful. What appears to be an absence of gratitude is often a well of other important and unacknowledged emotions, namely grief, disappointment, sadness, despair or fear. For children, it is often impossible to set these emotions aside to show appreciation. For adults, it is also difficult, and yet we try to squelch these feelings in ourselves and even in others. Consider that grief is not the opposite of gratitude; it may be the necessary emotion to move through to access gratitude underneath.

## Make Room for Both

I believe genuine gratitude always comes with an acknowledgement of some form of grief — of the ways things are not, the ways things never were or never will be, or of what and whom are missing. And believe it or not, making space for that grief can actually increase our capacity to embrace and appreciate what *is* here right now. So, this season, I invite you to hold gratitude in one hand, even as you hold grief in the other. After all, you have two hands for a reason. You can hold both.

## Reference

1. Jans-Beken, L, Jacobs, N., Janssens, M, Peeters, S, Reijnders, J, Lechner, L, and Lataster, J. Gratitude and Health: An Updated Review. *The Journal of Positive Psychology*, 15(6), 743–782. Accessed at www.tandfonline.com/doi/full/10.1080/17439760.2019.1651888.

**MAIREAD MCCONNELL,** PhD, is a clinical psychologist and assistant professor at Banner University Medical Center in Tucson, Ariz. She specializes in health psychology and is passionate about helping patients live well while navigating the challenges of chronic illness.

# Reconnect with volunteer days

People with primary immunodeficiency (PI) who infuse CUVITRU weekly or every other week may be able to experience more of these moments.

## What is CUVITRU®?

CUVITRU [Immune Globulin Subcutaneous (Human)] 20% Solution is a ready-to-use liquid medicine that is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years and older.

## IMPORTANT SAFETY INFORMATION

**What is the most important information I need to know about CUVITRU?**

CUVITRU can cause the following serious reactions:

- Severe allergic reactions causing difficulty in breathing or skin rashes
- Decreased kidney function or kidney failure
- Blood clots in the heart, brain, lungs, or elsewhere in the body
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting
- Dark colored urine, swelling, fatigue, or difficulty breathing

**Who should not use CUVITRU?**

Do not use CUVITRU if you:

- Have had a severe allergic reaction to immune globulin or other blood products.
- Have a condition called selective (or severe) immunoglobulin A (IgA) deficiency.

**What should I avoid while taking CUVITRU?**

- CUVITRU can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your healthcare provider (HCP) that you take CUVITRU.
- Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing.

**What are the possible or reasonably likely side effects of CUVITRU?**

**CUVITRU can cause serious side effects. If any of the following problems occur after starting CUVITRU, stop the infusion immediately and contact your HCP or call emergency services:**

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot.

Cuvitru
[Immune Globulin Subcutaneous (Human)] 20%

## Proven protection from infection

In the North American (NA) study, there were 0.012 acute serious bacterial infections (ASBIs) per patient-year.*† This exceeds the FDA standard for effectiveness, which is one serious ASBI per year.

## Nearly all infusions (99.8%) were completed without reduction, interruption or discontinuation due to tolerability

No patients discontinued due to local adverse reactions (ARs) and 0 serious ARs related to CUVITRU were reported.

The most common adverse reactions observed in clinical trials in ≥5% of patients were: local adverse reactions including mild or moderate pain, erythema, and pruritus, and systemic adverse reactions including headache, nausea, fatigue, diarrhea, and vomiting.

## Flexible administration that can be tailored to fit your lifestyle‡§

CUVITRU can be infused at the fastest rates and highest volumes with the fewest infusion sites of any subQ IG.§

In the NA clinical study, CUVITRU was studied in 77 people with PI ≥2 years of age. The main goal of the study was to measure how many acute serious bacterial infections (ASBIs) were experienced over the course of 1 year. ASBIs are short-term but serious infections that require immediate medical care. ASBIs were evaluated in 74 people taking CUVITRU for an average of 380.5 days (range, 30-629 days).

*One ASBI that occurred during the study was a case of pneumonia in a 78-year-old person.

†A patient-year is a patient experience in a clinical trial over the course of 1 year. One patient-year is equal to, for example, the experience of 2 patients for 6 months, or 12 patients for 1 month each.

‡In the NA study, the average infusion time was 0.95 hours (range 0.2-6.4 hours) and most (84.9%) used 1 to 2 needlesticks.

§You'll infuse your first 2 infusions at 10 to 20 mL/hr/site. After that, you'll be able to increase your rate up to 60 mL/hr/site as tolerated. Infuse at up to 4 sites simultaneously.

SubQ IG=subcutaneous immune globulin.

## IMPORTANT SAFETY INFORMATION (continued)

- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be sign of an infection.

The following one or more possible side effects may occur at the site of infusion. These generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Itching

The most common side effects that may occur are:

- Headache
- Nausea
- Fatigue
- Diarrhea
- Vomiting

**These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.**

***Please see Important Facts about CUVITRU on the following page.***

**You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.**

**Up to 100% of out-of-pocket co-pay costs could be covered.**

Scan the QR code to learn more about CUVITRU, including co-pay costs.



Takeda

## IMPORTANT FACTS about CUVITRU (CUE-vih-troo) [Immune Globulin Subcutaneous (Human)] 20% Solution

### What is the most important information I need to know about CUVITRU?

CUVITRU can cause the following serious reactions:

- Severe allergic reactions causing difficulty in breathing or skin rashes
- Decreased kidney function or kidney failure
- Blood clots in the heart, brain, lungs, or elsewhere in the body
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting
- Dark colored urine, swelling, fatigue, or difficulty breathing

### What is CUVITRU?

CUVITRU is a ready-to-use liquid medicine that contains immunoglobulin G (IgG) antibodies, which protect the body against infection. CUVITRU is used to treat patients with primary immunodeficiency diseases (PI).

There are many forms of PI. The most common types of PI result in an inability to make a very important type of protein called antibodies, which help the body fight off infections from bacteria or viruses. CUVITRU is made from human plasma that is donated by healthy people. CUVITRU contains antibodies collected from these healthy people that replace the missing antibodies in PI patients.

### Who should not use CUVITRU?

Do not use CUVITRU if you have a known history of a severe allergic reaction to immune globulin or other blood products. If you have such a history, discuss this with your healthcare provider (HCP) to determine if CUVITRU can be given to you. Tell your HCP if you have a condition called selective (or severe) immunoglobulin A (IgA) deficiency.

### How should I use CUVITRU?

CUVITRU is given under the skin (subcutaneously). Most of the time, infusions under the skin are given at home by self-infusion or by caregivers. Instructions for giving CUVITRU under the skin (subcutaneously) are provided in the FDA-approved patient labeling (Information for Patients and Instructions for Use). Only use CUVITRU by yourself after you have been instructed by your HCP.

### What should I avoid while taking CUVITRU?

CUVITRU can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your HCP that you take CUVITRU.

Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing.

### What are the possible or reasonably likely side effects of CUVITRU?

The following are one or more possible reactions that may occur at the site of infusion. These generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Itching

The most common side effects of CUVITRU are headache, nausea, fatigue, diarrhea, and vomiting.

If any of the following problems occur after starting treatment with CUVITRU, stop the infusion immediately and contact your HCP or call emergency services. These could be signs of a serious problem.

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be a sign of an infection.

These are not all the possible side effects. You can ask your HCP for a physician's information leaflet. Tell your HCP about any side effect that bothers you or that does not go away.

Whenever giving yourself treatments at home, you should have another responsible person present to help treat side effects or get help if you have a serious adverse reaction occur. Ask your HCP whether you should have rescue medications, such as antihistamines or epinephrine.

### How do I store CUVITRU?

Store CUVITRU refrigerated or at room temperature.

- You can store CUVITRU in the refrigerator (36°F to 46°F [2°C to 8°C]) for up to 36 months or
- You can store CUVITRU at room temperature (up to 77°F [25°C]) for up to 24 months.
- Do not return CUVITRU to the refrigerator if you take it out to room temperature.
- Do not freeze.
- Do not shake.
- Check the expiration date on the carton and vial label. Do not use CUVITRU after the expiration date.
- Protect from light. You can use the original CUVITRU containers to protect it from light.

### How do I get more information about CUVITRU?

The risk information provided here is not comprehensive. To learn more, talk about CUVITRU with your HCP or pharmacist. The FDA-approved Full Prescribing Information, including Information for Patients, can be found at www.CUVITRU.com or by calling 1-877-TAKEDA7 (1-877-825-3327).



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US-CUV-0408v1.0 07/21

# SARS-CoV-2 and COVID-19: Why Does Our Immune System Have a Problem with This Virus? Part 3

By Terry O. Harville, MD, PhD

**PREVIOUSLY, WE** discussed how the immune system can have difficulty dealing with zoonotic viruses such as influenza, HIV and, in particular, SARS-CoV-2 (SARS2). In this column, we will further discuss details of the immunologic concepts.

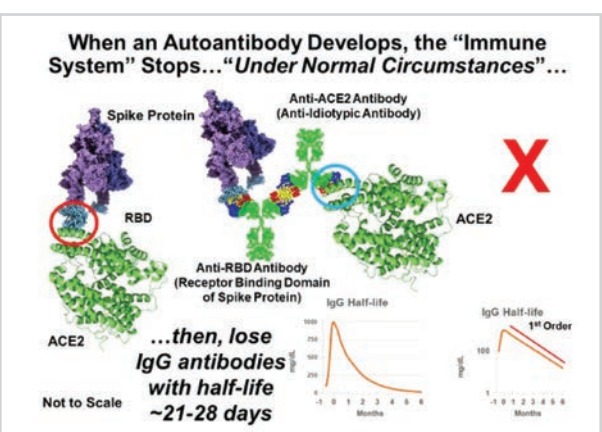
SARS2 binds to cells through the receptor binding domain (RBD) region of its spike protein to the ACE2 protein on cell surfaces for entering into and infecting cells. Thus, the RBD is an important structure with regard to immune system activity toward SARS2. The so-called "neutralizing" vaccines use this region, or an extended spike protein encompassing this region, to generate an antibody against the RBD, and then bind to the RBD, so there will be no available structure to bind to ACE2. Therefore, the antibody blocks binding to ACE2, which in turn prevents the virus from entering into and infecting the cell. Therefore, it becomes a "neutralizing antibody."

This process also occurs with natural infection. Until we studied and published articles on this, others failed to consider the Jerne Network Theory of the Immune System.[1,2,3] This is when an antibody forms to a specific target, in this case the RBD of SARS2, making the antibody a new protein that can be a target for new antibody production. The antibody against this RBD target is termed an "idiotype," and the antibody directed to the anti-RBD antibody is termed and "anti-idiotype." Thus, there is an "idiotype and an anti-idiotype" interaction. Since the RBD target is ACE2, the idiotypic antibody to RBD has molecular features that make it appear to be ACE2. As a consequence, the anti-idiotypic antibody binds to ACE2, which is bad because this can disrupt ACE2 function. We demonstrated this in our studies as shown in the figure.

Further, since the production of anti-ACE2 antibodies is in essence "auto-immune" antibody production, the normally functioning immune system shuts down this process (see the red X in the figure). This is why we then lose the antibodies in a predictable fashion and need revaccinations. To maintain high levels of anti-SARS2, the anti-RBD neutralizing antibody requires vaccination every three to six months. For practical reasons, annual vaccinations or even twice yearly vaccines may be needed for the foreseeable future. The good news is that vaccination has demonstrated very good and lasting T lymphocyte immunity. Thus, SARS2 vaccination is not to prevent infection (antibodies lost too soon), but to prevent severe illness and death in immunocompetent persons. Overall, this illustrates why herd immunity will likely never be achieved (antibodies cannot persist long enough). And, most importantly, this is why persons who are immunocompromised or have an immunodeficiency will remain at risk. "Wear your mask!"

In the next column, I will discuss more about SARS2.

**Figure. SARS-CoV-2 Antibody and Autoantibody Production**




*The RBD (receptor binding domain) (blue in the red circle) of the spike protein (purple) binds to ACE2 (green coils) (left side of the figure). Natural infection or vaccination induces formation of anti-RBD antibody (middle left of figure). This induces the formation of the anti-idiotypic, anti-ACE2 antibody (top middle of figure), which can bind to ACE2 (middle right of figure, light blue circle). Under typical circumstances, with an otherwise normal functioning immune system, this antibody production is detected as autoantibody, which then shuts down the production (red X). Antibodies are then lost at a predictable rate, where after approximately six months, little protective antibody immunity remains (lower middle and lower right of figure).*

## References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3. Harville, TO, and Arthur, JM. Anti-Idiotypic Antibodies in SARS-CoV-2 Infection and Vaccination. *The New England Journal of Medicine*, 386(5): Feb. 2, 2022 Accessed at www.nejm.org/doi/full/10.1056/NEJMc2119443.


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# Understanding Venous Access for IVIG Infusions

By Michelle Greer, RN, IgCN

**VENOUS ACCESS** is an important consideration when creating an immune globulin (IG) treatment plan. And, since most patients who receive intravenous IG (IVIG) use their venous access solely for this therapy, a decision should be made if access should be peripheral or central.

## Treatment Considerations

When a physician prescribes IG, the dose, frequency and length of therapy *can*, but does not *always*, dictate the type of venous access to be used. The patient should have input based on lifestyle and personal preferences, but the physician must first determine clinically what the best options are for the patient. In addition, insurance companies may narrow the patient's options.

For instance, when deciding on the route of administration (either intravenously or subcutaneously), some health plans require members to have a clinical reason for approving subcutaneous IG (SCIG), one of which is poor venous access. Another accepted justification is if the patient has had previous tolerability issues with IVIG. Lifestyle preference, however, is not accepted as a reason to approve SCIG.

Determining site of care for IVIG infusions (at home, in a physician's office or in a hospital outpatient or freestanding infusion center) should also be up to the patient and physician. The home setting offers convenience and comfort, as well as more scheduling flexibility than outpatient settings, but the physician may prefer to infuse in his or her own office or in an affiliated hospital infusion center for other reasons, one of which can be tolerability issues with the infusions. Conversely, since home infusions are typically the most cost-effective site of care, health plans might lean toward this setting as a way to save money.

Venous access is also a crucial treatment consideration. A decision must be made about whether the infusions should be given via peripheral IV access that is established for each infusion, or whether a central line should be placed. This is often determined by the length of time a patient will need therapy. For some patients, IG can be a lifetime therapy, particularly for those with a primary immune deficiency (PI) or an autoimmune condition. For others, it can be long-term, but not for life. And, in some instances, the length of treatment is not known at the time treatment is initiated, and treatment frequency can vary depending on the prescriber and the condition being treated.

## Venous Access Challenges

In most cases, establishing peripheral IV access is not problematic, and there is no venous access consideration in between infusions since access is discontinued after each infusion is completed. There may be times, however, when venous access *is* challenging.

When IVIG is prescribed, regardless of site of care, a nurse who is skilled at peripheral IV placement will perform the procedure. While some people don't have easily accessed veins (sometimes due to a person's anatomy or sometimes due to long-term IV use that can damage the vein and cause scar tissue to form permanently), nurses who are adept at IV placement can successfully secure access without an issue even in tough situations. Most institutions and home infusion providers will have a policy and procedure in place regarding peripheral IV placement, including number of attempts to secure access. Three access attempts is the typical maximum; however, that may be extended with patient permission. At times, another nurse might be called in if the situation and/or site of care allows for it. Additionally, vein location devices are sometimes available and utilized in challenging situations.

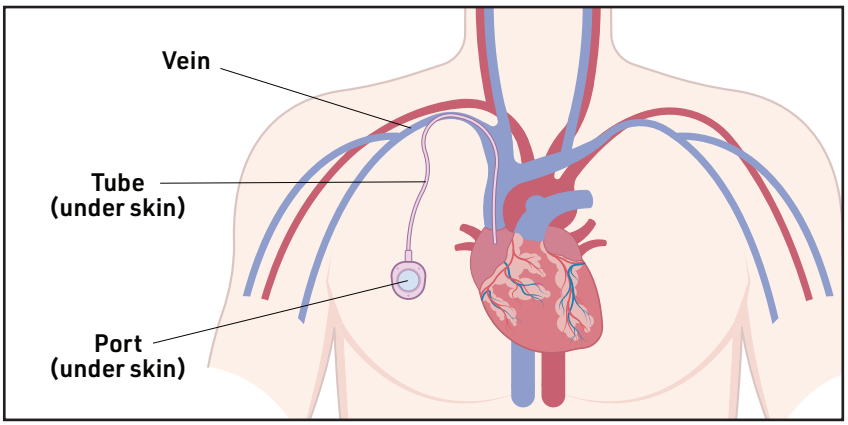
## Resolving Venous Access Challenges

If there are ongoing venous access challenges, they must be resolved. Depending on the IG dosage and patient preference, switching to subcutaneous infusions might make the most sense. However, if the dose is too high or if the person does not want to or cannot self-administer treatment, a central venous catheter, commonly called a central line, might be the solution.

There are various types of central catheters, and not all are appropriate for an IVIG patient. A central line's insertion point can be in the arm or chest, but the internal tip of the catheter is always in a larger, more central blood vessel. This can be the subclavian vein or the superior vena cava. Although some central lines might be placed in the neck for the jugular vein or in a location to infuse into a femoral vein, these lines would never be used for IVIG infusions. Infusing any therapy into these large vessels aids in dilution of the treatment. In addition to infusing any medication into a central line, blood can also be taken from a central line for lab work, eliminating the need for another needle stick.

There are two particular types of central lines that might be used for IVIG infusions. The first is a port. Placement of a port is usually an outpatient procedure

**Vascular Access Port Implantation**




performed by a physician. It is a short and minor surgery to insert the port under the skin in the upper chest, with the catheter placed into the blood vessel. Prior to an infusion, a special needle is used to access the port, and once the infusion is complete, the needle is removed; there is nothing on the outside of the chest in between infusions since there is a thin layer of skin covering the port. When not in use, a port usually needs to be flushed monthly to keep it unobstructed. However, this is usually not an issue with IVIG since infusions are typically administered at least monthly.

The other type of central line that might be used for IVIG infusions is a peripherally inserted central catheter, or PICC. A PICC can be placed by a specially trained nurse in any setting, or it can be placed by a physician in an outpatient setting. A PICC is placed in the upper arm and fed through a vein until the tip is properly placed in the subclavian vein or the superior vena cava. Confirmation of proper placement is confirmed by an X-ray or ultrasound.

A PICC is generally less desirable for IVIG treatment for several reasons. First, there is an external component to the catheter that needs to be cared for on a more frequent basis. Second, while there is no consideration for showering or swimming with a port since there is nothing outside of the body, a PICC needs to be covered securely so it won't get wet. Lastly, the risk for a PICC to be dislodged is greater than with a port. One difference between a PICC and a port is that a PICC can stay in place for a shorter duration.

## Complications of Central Lines

All central lines have potential complications that need to be considered to determine which is right for someone receiving IVIG. Mechanical issues are one complication, mainly an occluded line. While care is taken to flush out the line after an infusion or blood draw, a clot in the line can develop. Fortunately, measures can be taken to quickly dissolve the clot.

Risk of a blood clot outside of the catheter in a blood vessel is also an important consideration. All brands of IVIG include the same boxed warning of adverse events, including thrombosis. However, each individual's risk factors, however small, that increase the risk of a thrombosis must be assessed. This includes previous thrombotic events such as myocardial infarction or stroke, age, mobility, hypercoagulable conditions and other medications that increase the risk of clotting.

Another potential complication of a central line is infection. Because a central line is an implanted device, there is the risk of infection at the insertion site. While the possibility of bacteria getting into the bloodstream and causing an infection inside the body is low, it *can* happen. And for someone with a PI, this could be especially problematic. There is some thought within the immune deficiency community that a central line should never be used because SCIG is a better alternative.

The American Academy of Allergy, Asthma and Immunology (AAAAI) published a practice parameter for the diagnosis and management of PI. AAAAI's statement 14 states, "The placement of permanent central venous access solely for the purpose of IVIG administration should be discouraged.… Permanent central venous catheters can be associated with thrombotic and infectious complications. For patients who require intravenous access only for IgG administration every 2 to 4 weeks, permanent indwelling catheters might not represent an acceptable risk. Difficult venous access need not be a compelling indication for catheter placement with the growing availability of subcutaneous IgG infusion."

## Venous Access Is Vital

All in all, proper venous access is vital to successful IVIG infusions. When peripheral venous access becomes a potential obstacle to a successful infusion, a central line might be a viable alternative. SCIG might also be a good solution. The patient and doctor should discuss and decide together what is best for managing the infusions and the condition being treated.

**MICHELLE GREER**, RN, IgCN, is senior vice president of sales at Nufactor, a specialty infusion company.

MEDICINES

# FDA Approves Yimmugo IVIG Product to Treat PI; Kedrion to Distribute

Biotest AG has received approval from the U.S. Food and Drug Administration (FDA) for Yimmugo (human — dira, 10% liquid), a newly developed polyvalent sugar-free intravenous immune globulin (IVIG) therapeutic, to treat primary immunodeficiencies (PI) in patients 2 years and older. Kedrion Biopharma Inc. announced that it has established the framework for a long-term agreement with Biotest AG for the full commercialization and distribution of Yimmugo in the U.S.

Approval of Yimmugo was based on data from an open-label, prospective, Phase III study that evaluated the efficacy and safety of Yimmugo in 67 patients (mean age 35 years) with PI, who had established IVIG therapy for at least three months with a constant dose, and at least one IgG trough level of at least 5g/L during the previous three months.

Study participants received Yimmugo 0.2 to 0.8g/kg body weight at either every three or four weeks for 12 months. The primary endpoint was the rate of serious bacterial infections (SBI), defined as bacterial pneumonia, bacteremia/septicemia, osteomyelitis/septic arthritis, visceral abscesses or bacterial meningitis over a period of 12 months.

The acute SBI rate was reported to be 0.07 during the 12-month study period, which met the predefined success rate of less than one acute SBI per patient per year. Five patients experienced an acute SBI (all five episodes were bacterial pneumonia). The most common adverse reactions observed with treatment were headache, upper respiratory tract infections, fatigue, nausea and increased blood pressure.

Yimmugo is supplied in a single-dose vial as a solution containing 10% immunoglobulin G (100mg/mL) in the following presentations: 5g in 50mL; 10g in 100mL; 20g in 200mL. The product is expected to be available in the second half of 2024. IG

Park, B. FDA Approves Yimmugo for Primary Humoral Immunodeficiency. Medical Professionals Reference, June 17, 2024. Accessed at www.empr.com/home/news/fda-approves-yimmugo-for-primary-humoral-immunodeficiency.

Kedrion Biopharma to Distribute Biotest's Immunoglobulin Therapy Yimmugo in US Market. Kedrion Biopharma press release, July 2, 2024. Accesesd at www.pharmabiz.com/NewsDetails.aspx?aid=170071&sid=2.

MEDICINES

# FDA Approves XOLREMDI to Treat WHIM Syndrome

The U.S. Food and Drug Administration (FDA) has approved X4 Pharmaceuticals' XOLREMDI (mavorixafor) capsules for use in patients 12 years of age and older with WHIM (warts, hypogammaglobulinemia, infections and myelokathexis) syndrome to increase the number of circulating mature neutrophils and lymphocytes. XOLREMDI, a selective CXC chemokine receptor 4 (CXCR4) antagonist, is the first therapy specifically indicated in patients with WHIM syndrome, a rare, combined primary immunodeficiency and chronic neutropenic disorder caused by CXCR4 pathway dysfunction.

FDA approval of XOLREMDI was based on results of the pivotal, 4WHIM Phase III clinical trial, a global, randomized, double-blind, placebo-controlled, 52-week multicenter study that evaluated the efficacy and safety of XOLREMDI in 31 people aged 12 years and older diagnosed with WHIM syndrome. The efficacy of XOLREMDI was determined by improvement in absolute neutrophil counts (ANC), improvement in absolute lymphocyte counts (ALC) and a reduction in infections. In the 4WHIM trial, XOLREMDI treatment demonstrated increased time above threshold (≥500 cells/microliter) for ANC vs. placebo and increased time above threshold (≥1000 cells/microliter) for ALC vs. placebo. The efficacy of XOLREMDI was further assessed in a composite endpoint consisting of total infection score and total wart change score using a Win-Ratio method. Analyses of the individual components of this composite endpoint showed an approximate 40 percent reduction in total infection score, weighted by infection severity, in XOLREMDI-treated patients compared with placebo-treated patients. There was no difference in total wart change scores between the XOLREMDI and placebo treatment arms over the 52-week period. Treatment with XOLREMDI also resulted in a 60 percent reduction in the annualized infection rate compared with placebo-treated patients. IG

X4 Pharmaceuticals Announces FDA Approval of XOLREMDI™ (mavorixafor) Capsules, First Drug Indicated in Patients with WHIM Syndrome. BioSpace, April 29, 2024. Accessed at www.biospace.com/article/releases/x4-pharmaceuticals-announces-fda-approval-of-xolremdi-mavorixafor-capsules-first-drug-indicated-in-patients-with-whim-syndrome.

RESEARCH

# Study Finds IEI Patients Have Higher Vaccination Rates Against COVID-19 Than Other Respiratory Tract Infection Vaccines

A recent study that examined the vaccination and respiratory tract infection rates in a diverse inborn errors of immunity (IEI) patient cohort undergoing immune globulin replacement therapy (IGRT) found there were higher vaccination rates for the COVID-19 vaccine compared to the influenza and pneumococcal vaccines.

In the study, the researchers retrospectively evaluated 33 IEI patients (mean age= 37.7 ± 11.4 years; 17 male) on IGRT at a tertiary care center. Data on vaccinations and respiratory infections were extracted from medical records. The most common clinical phenotype in the cohort was primary antibody deficiencies (90.9 percent). Only two patients had a genetic diagnosis, both of whom were brothers diagnosed with Wiskott-Aldrich syndrome (WAS). Almost half (48.5 percent) of the patients had bronchiectasis and 81.8 percent were on prophylactic antibiotics. All patients with IEI included in the study were regularly receiving IGRT.

The vaccination rate of patients against respiratory tract infections was 42.4 percent, 57.6 percent and 78.8 percent for influenza, pneumococcus and COVID-19, respectively. Only one patient (7.1 percent) who received the influenza vaccine developed an upper respiratory tract infection. However, viral panel analysis could not be performed for this patient as he or she did not present to the hospital. The COVID-19 vaccination rate was notably higher than that of other vaccines, likely due to increased awareness during the pandemic, aided by public advisories and media influence.

The researchers concluded that although they observed the potential impact of social and governmental influence in increasing vaccination rates, it is crucial to acknowledge that vaccination decisions in IEI patients must be individualized.

Bayrak Durmaz, MS, Yildiz, R, Keskin, G, and Altner, S. Vaccination Against Respiratory Tract Pathogens in Primary Immune Deficiency Patients Receiving Immunoglobulin Replacement Therapy. *Tuberkuloz ve Toraks*, 2024;72(1):1-8. Accessed at avesis.ankara.edu.tr/yayin/fe6a21a4-3682-45cf-a657-d605d23802a5/vaccination-against-respiratory-tract-pathogens-in-primary-immune-deficiency-patients-receiving-immunoglobulin-replacement-therapy-primer-bagisiklik-yetmezligi-hastalarinda-solunum-yolu-patojenlerine.

DOCUMENTARY

# Wait List Debuts to Host Screening of IDF's PI Documentary

The Immune Deficiency Foundation (IDF) is accepting waitlist entries for those who would like to host a screening of the organization's anticipated documentary "Compromised: Life Without Immunity." IDF held a special screening of the documentary at its 2024 PI Conference held June 20-22 in Chicago. The film, directed and created by staff member Zachary Moore, depicts the lives of several primary immunodeficiency (PI) community members as they recount their journeys to diagnosis, explain their treatments and discuss how PI has impacted them and their families.

The documentary features Ben Everett, 11, diagnosed with common variable immunodeficiency (CVID), and his parents, Kim Everett, also diagnosed with PI, and Matt Everett, of Los Angeles, Calif.; Dionne Stalling, 54, of St. Louis, Mo., who is diagnosed with CVID and founded Rare and Black; Victoria Medl, 20, of New Jersey, who has an unspecified PI and is a student at Rutgers University, and her mother, Agnes Medl; Shane Brisson, 54, diagnosed with chronic granulomatous disease, who works as a magazine creative director and lives in St. Petersburg, Fla.; Jerry Shorten, 37, diagnosed with severe combined immunodeficiency (SCID), who is a residential developer in Houston, Texas; and Yvette Shorten, mother to Jerry and two other sons, both of whom passed away from complications of SCID.

While each person's story is different, the common themes of determination, longing, acceptance, joy and hope carry viewers through the PI experience as the individuals interviewed describe their lives.

Interspersed between the five segments of patients sharing their stories are healthcare providers who discuss the hardships PI patients face such as time to diagnosis, barriers to treatment and co-morbidities due to PI.

To sign up for the waitlist, go to primaryimmune.org/resources/news-articles/compromised-life-without-immunity-profiles-families-affected-pi-0.

'Compromised: Life Without Immunity' Profiles Families Affected by PI. Immune Deficiency Foundation news release, June 27, 2024. Accessed at primaryimmune.org/resources/news-articles/compromised-life-without-immunity-profiles-families-affected-pi-0.

RESEARCH

## Scientists Discover Defect That Causes Lupus

Northwestern Medicine and Brigham and Women's Hospital scientists have discovered a molecular defect that promotes the pathologic immune response in systemic lupus erythematosus (known as lupus) and that reversing this defect may potentially reverse the disease.

According to the scientists, there are disease-associated changes in multiple molecules in the blood of patients with lupus that ultimately lead to insufficient activation of a pathway controlled by the aryl hydrocarbon receptor (AHR), which regulates cells' response to environmental pollutants, bacteria or metabolites. Insufficient activation of AHR results in too many disease-promoting immune cells, called the T peripheral helper cells, that promote the production of disease-causing autoantibodies.

To show this discovery can be leveraged for treatments, the investigators returned the AHR-activating molecules to blood samples from lupus patients, which seemed to reprogram these lupus-causing cells into a cell called a Th22 cell that may promote wound healing from the damage caused by this autoimmune disease.

"We found that if we either activate the AHR pathway with small molecule activators or limit the pathologically excessive interferon in the blood, we can reduce the number of these disease-causing cells," said Jaehyuk Choi, MD, PhD, associate professor of dermatology at Northwestern University Feinberg School of Medicine and a Northwestern Medicine dermatologist. "If these effects are durable, this may be a potential cure."

The researchers next want to expand their efforts into developing novel treatments for lupus patients. They are now working to find ways to deliver these molecules safely and effectively to people. IG

Scientists Discover a Cause of Lupus and a Possible Way to Reverse It. Northwestern University, July 10, 2024. Accessed at www.eurekalert.org/news-releases/1050664.

RESEARCH

## Study Shows New Pill May Improve Muscle Strength in Myasthenia Gravis

A team of researchers from Aarhus University revealed a breakthrough in the treatment of myasthenia gravis (MG): a pill that improves communication between motor nerves and muscle fibers.

MG is an autoimmune disease in which the immune system attacks the connection between nerves and muscle cells that leads to severe muscle weakness and extreme fatigue. It can affect breathing and be life-threatening — symptoms researchers wanted to combat by strengthening the function of the contact point between nerves and muscle cells.

During his PhD project at Aarhus University, Thomas Holm Pedersen, associate professor at the Department of Biomedicine at Aarhus University, CEO of NMD Pharma and lead author of the study discovered that the so-called CIC-1 chloride channels at the neuromuscular junction are crucial for muscle activation. "This led to the idea of using the CI channels as a new treatment point for diseases where the neuromuscular connection is compromised, including myasthenia gravis," he explained.

Researchers demonstrated they could strengthen the muscles' ability to respond to nerve impulses by targeting a treatment at the specific channel, which they think may increase muscle strength and reduce fatigue in patients.

"We tested it on patients who had to take a tablet daily for their muscle weakness and fatigue, and we could see that the treatment concept worked," Dr. Pedersen said. "The patients became stronger."

The treatment was successful in increasing muscle strength among patients, but unlike existing treatments for MG, it may well prove to be free from side effects. "We won't know for sure for a few years, after we've conducted more studies, but it looks really promising right now," said Dr. Pedersen.

The breakthrough may significantly improve the quality of life for patients struggling with MG, as well as advance understanding and treatment of other neuromuscular diseases. "This study summarizes years of work here at Aarhus University and NMD Pharma. We have proven that the method works in patients, and now we continue with the clinical trials to bring the drug to the patients and to explore its broader application," he said.

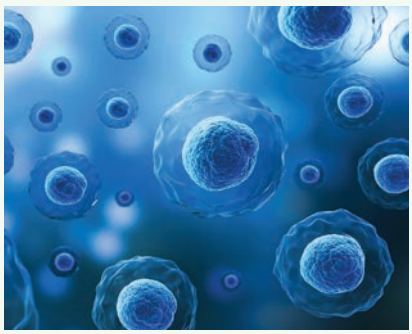
The research team is currently planning more follow-up clinical trials, including another study on MG and one on the hereditary disease spinal muscular atrophy, which leads to muscle wasting. IG

Christensen, JB. Study: Pill Makes Patients with Life-Threatening Muscle Weakness Stronger. Aarhus University, March 21, 2024. Accessed at dent.au.dk/en/display/artikel/study-pill-makes-patients-with-life-threatening-muscle-weakness-stronger.

RESEARCH

# FDA Green Lights Phase II Trial of Cell Therapy for SPS

The U.S. Food and Drug Administration (FDA) has granted Kyverna Therapeutics permission to begin clinical trials of its autologous anti-CD19 CAR T-cell therapy candidate KYV-101 for treatment-refractory stiff-person syndrome (SPS). This is the eighth investigational new drug (IND) application for KYV-101, Kyverna's lead candidate, that has been cleared by the FDA.



The new IND clearance from FDA enables Kyverna to begin an open-label Phase II clinical trial (KYSA-8) in which investigators will test KYV-101 in patients with SPS, a rare and progressive neurological autoimmune disorder that can lead to debilitating muscle stiffness. KYV-101 is already being clinically tested in other B cell-driven autoimmune indications, including lupus nephritis, systemic sclerosis and myasthenia gravis.

To date, 30 patients have been treated with KYV-101 across 15 rheumatological and neurological autoimmune conditions, and investigators have observed promising safety results and drug-free remission in most patients.

Kyverna Therapeutics to Launch Phase II Trial of Cell Therapy in Stiff-Person Syndrome. Precision Medicine Online, June 20, 2024. Accessed at www.precisionmedicineonline.com/regulatory-news-fda-approvals/kyverna-therapeutics-launch-phase-ii-trial-cell-therapy-stiff-person.

RESEARCH

# Study of Mezagitamab Demonstrates Potential to Transform ITP Treatment

Results from Takeda's Phase IIb, randomized, double-blind, placebo-controlled study evaluating the safety, tolerability and efficacy of mezagitamab in patients with persistent or chronic primary immune thrombocytopenia (ITP) showed it has a favorable safety/tolerability profile. ITP, a rare immune-mediated bleeding disorder, is characterized by the accelerated destruction of platelets in blood, resulting in a decreased platelet count and an increase of bleeding that can be debilitating.

The trial evaluated three different doses of subcutaneous mezagitamab (100 mg, 300 mg and 600 mg) versus placebo, given once weekly for eight weeks in patients with chronic or persistent primary ITP, followed by more than eight weeks of safety follow-up. The primary endpoint was the percentage of patients with at least one grade 3 or higher treatment emergent adverse events, serious adverse events and adverse events (AEs) leading to mezagitamab discontinuation. Secondary endpoints included platelet response, complete platelet response, clinically meaningful platelet response and hemostatic platelet response.

Trial results demonstrated that mezagitamab treatment considerably improved platelet response compared to placebo across all three dose levels of mezagitamab tested. Patients treated with mezagitamab showed rapid and sustained increases in platelet counts (above the 50,000/μL therapeutic threshold) that persisted eight weeks after the last dose through to week 16, illustrating the rapid and post-therapy effects of mezagitamab on platelet response.

All the different measures of platelet response evaluated were highest among patients treated with the mezagitamab 600 mg dose. Specifically, 81.8 percent achieved complete platelet response, 90.9 percent achieved clinically meaningful platelet response and 100 percent achieved hemostatic platelet response.

Fewer mezagitamab-treated patients compared to placebo had ≥1 disease activity-related bleeding AE (17.9 percent vs. 46.2 percent, respectively).

"Despite treatment with currently available therapies, there is still a significant disease burden and need for a disease-modifying treatment that people living with ITP can tolerate," said David Kuter, MD, PhD, a leading expert in ITP. "These Phase IIb trial results are especially encouraging because they show mezagitamab's favorable efficacy and safety profile — setting the stage for the generation of additional clinical evidence for this anti-CD38 monoclonal antibody with best-in-class potential for efficacy in ITP."

Takeda Presents Late-Breaking Data from Phase 2b Study of Mezagitamab, Demonstrating Potential to Transform Treatment of Primary Immune Thrombocytopenia. Takeda news release, June 24, 2024. Accessed at www.takeda.com/newsroom/newsreleases/2024/late-breaking-data-from-phase-2b-study-of-mezagitamab.

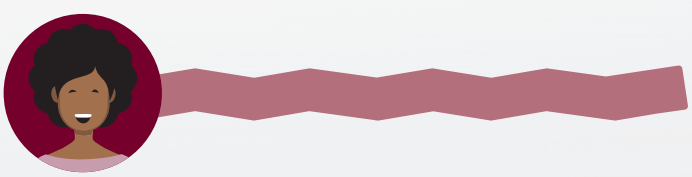
Hizentra®
Immune Globulin Subcutaneous (Human) 20% Liquid

PREFILLED SYRINGES

# Are ups and downs in your Ig levels affecting you?

## Hizentra helps you maintain steady Ig levels, which can make a difference in how you feel

**With IVIg dosing every 3–4 weeks**, you may experience highs and lows between infusions.



**Hizentra flexible dosing** provides smaller, more frequent doses than IVIg. More steady Ig levels may reduce the potential for side effects between infusions.

### Important Safety Information

Hizentra®, Immune Globulin Subcutaneous (Human), 20% Liquid, is a prescription medicine used to treat:

- Primary immune deficiency (PI) in patients 2 years and older
- Chronic inflammatory demyelinating polyneuropathy (CIDP) in adults

**WARNING: Thrombosis (blood clots) can occur with immune globulin products, including Hizentra. Risk factors can include: advanced age, prolonged immobilization, a history of blood clotting or hyperviscosity (blood thickness), use of estrogens, installed vascular catheters, and cardiovascular risk factors.**

**If you are at high risk of blood clots, your doctor will prescribe Hizentra at the minimum dose and infusion rate practicable and will monitor for signs of clotting events and hyperviscosity. Always drink sufficient fluids before infusing Hizentra.**

**See your doctor for a full explanation, and the full prescribing information for complete boxed warning.**

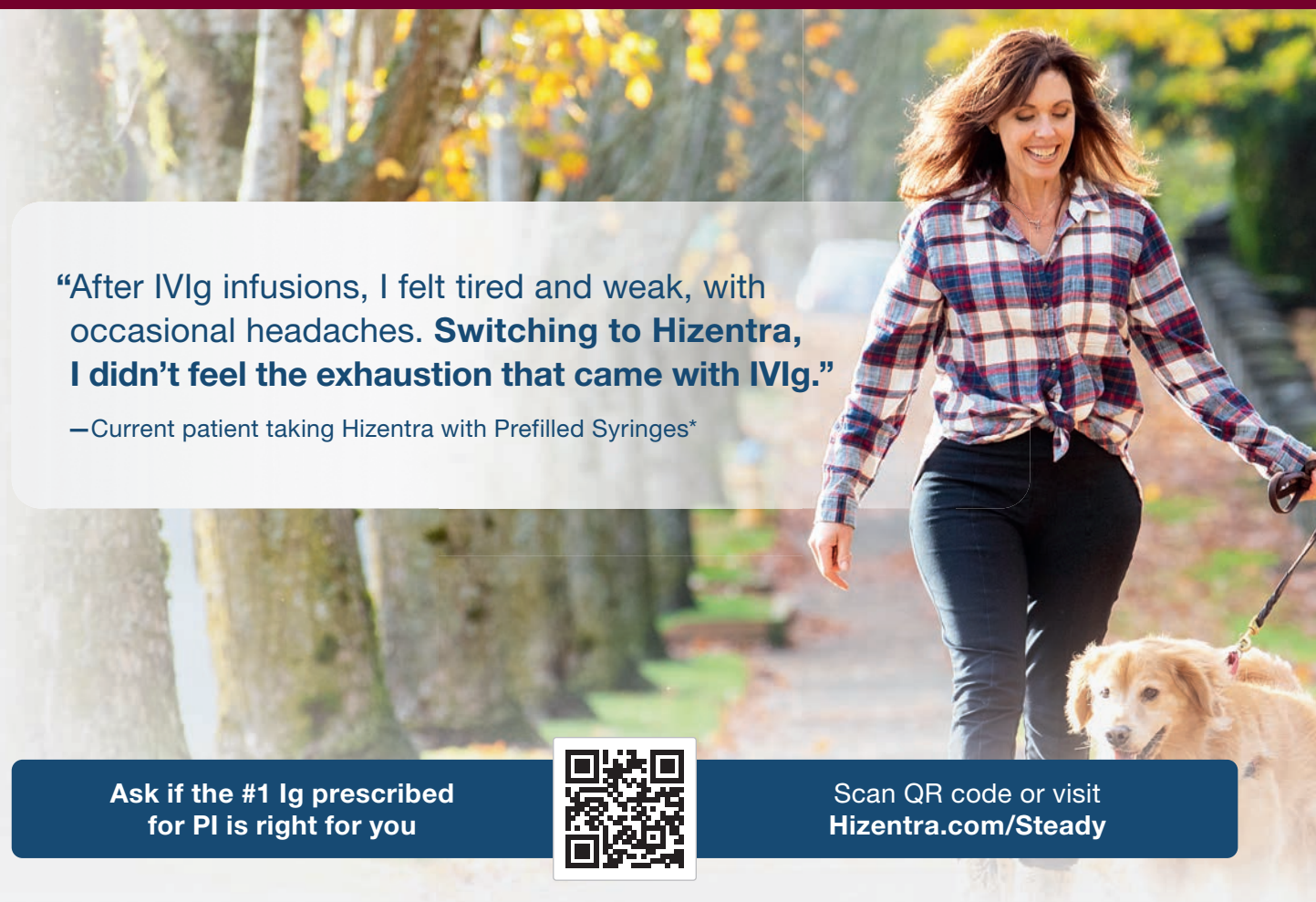
Treatment with Hizentra might not be possible if your doctor determines you have hyperprolinemia (too much proline in the blood), or are IgA-deficient with antibodies to IgA and a history of hypersensitivity. Tell your doctor if you have previously had a severe allergic reaction (including anaphylaxis) to the administration of human immune globulin. Tell your doctor right away or go to the emergency room if you have hives, trouble breathing, wheezing, dizziness, or fainting. These could be signs of a bad allergic reaction.

Inform your doctor of any medications you are taking, as well as any medical conditions you may have had, especially if you have a history of diseases related to the heart or blood vessels, or have been immobile for some time. Inform your physician if you are pregnant or nursing, or plan to become pregnant.

**Infuse Hizentra under your skin *only*; do not inject into a blood vessel.** Self-administer Hizentra only after having been taught to do so by your doctor or other healthcare professional, and having received dosing instructions for treating your condition.

Immediately report to your physician any of the following

**Please see Brief Summary of full Prescribing Information on reverse.**




Ig=immune globulin. IVIg=intravenous Ig. PI=primary immunodeficiency.
*Photo does not depict actual patient.

symptoms, which could be signs of serious adverse reactions to Hizentra:

- Reduced urination, sudden weight gain, or swelling in your legs (possible signs of a kidney problem).
- Pain and/or swelling or discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness/weakness on one side of the body (possible signs of a blood clot).
- Bad headache with nausea; vomiting; stiff neck; fever; and sensitivity to light (possible signs of meningitis).
- Brown or red urine; rapid heart rate; yellowing of the skin or eyes; chest pains or breathing trouble; fever over 100°F (possible symptoms of other conditions that require prompt treatment).

Hizentra is made from human blood. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent and its variant (vCJD), cannot be completely eliminated.

The most common side effects in the clinical trials for Hizentra include redness, swelling, itching, and/or bruising at the infusion site; headache; chest, joint or back pain; diarrhea; tiredness; cough; rash; itching; fever, nausea, and vomiting. These are not the only side effects possible. Tell your doctor about any side effect that bothers you or does not go away.

Before receiving any vaccine, tell immunizing physician if you have had recent therapy with Hizentra, as effectiveness of the vaccine could be compromised.

**Please see full prescribing information for Hizentra, including boxed warning and the patient product information.**

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.

You can also report side effects to CSL Behring's Pharmacovigilance Department at 1-866-915-6958.

CSL Behring

Hizentra®
Immune Globulin Subcutaneous (Human) 20% Liquid

PREFILLED SYRINGES

# Explore proven protection against infection in a convenient self-infused Ig

## Choose when and where you infuse*

SCIg with longest record of safety & effectiveness

Flexibility to fit your lifestyle


Steady Ig levels may improve how you feel vs IVIg

Simplify your infusions with prefilled syringes

**Ask if the #1 Ig prescribed for PI is right for you**

Scan QR code or visit **Hizentra.com/Steady**

SCIg=subcutaneous Ig. *After discussing with your doctor.

**HIZENTRA®, Immune Globulin Subcutaneous (Human), 20% Liquid**
**Initial US Approval: 2010**

**BRIEF SUMMARY OF PRESCRIBING INFORMATION**
**These highlights do not include all the information needed to use HIZENTRA safely and effectively. Please see full prescribing information for HIZENTRA, which has a section with information directed specifically to patients.**

**What is HIZENTRA?**
HIZENTRA is a prescription medicine used to treat primary immune deficiency (PI) and chronic inflammatory demyelinating polyneuropathy (CIDP). Infuse HIZENTRA only after you have been trained by your doctor or healthcare professional. HIZENTRA is to be infused under your skin only. DO NOT inject HIZENTRA into a blood vessel (vein or artery).

**Who should NOT take HIZENTRA?**
Do not take HIZENTRA if you have too much proline in your blood (called "hyperprolinemia") or if you have had reactions to polysorbate 80. Tell your doctor if you have had a serious reaction to other immune globulin medicines or have been told that you have a deficiency of the immunoglobulin called IgA.

Tell your doctor if you have a history of heart or blood vessel disease or blood clots, have thick blood, or have been immobile for some time. These things may increase your risk of having a blood clot after using HIZENTRA. Also tell your doctor what drugs you are using, as some drugs, such as those that contain the hormone estrogen (for example, birth control pills), may increase your risk of developing a blood clot.

**What are possible side effects of HIZENTRA?**
The most common side effects with HIZENTRA are:

- Redness, swelling, itching, and/or bruising at the infusion site
- Headache/migraine
- Nausea and/or vomiting
- Pain (including pain in the chest, back, joints, arms, legs)
- Fatigue
- Diarrhea
- Stomach ache/bloating
- Cough, cold or flu symptoms
- Rash (including hives)
- Itching
- Fever and/or chills
- Shortness of breath
- Dizziness
- Fall
- Runny or stuffy nose

Tell your doctor right away or go to the emergency room if you have hives, trouble breathing, wheezing, dizziness, or fainting. These could be signs of a bad allergic reaction.

Tell your doctor right away if you have any of the following symptoms. They could be signs of a serious problem.

- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain and/or swelling of an arm or leg with warmth over the affected area, discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, or numbness or weakness on one side of the body. These could be signs of a blood clot.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of a brain swelling called meningitis.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a blood problem.
- Chest pains or trouble breathing.
- Fever over 100°F. This could be a sign of an infection.

Tell your doctor about any side effects that concern you. You can ask your doctor to give you more information that is available to healthcare professionals.

**Please see full prescribing information, including full boxed warning and FDA-approved patient product information. For more information, visit Hizentra.com.**

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit *www.fda.gov/medwatch*, or call 1-800-FDA-1088.

You can also report side effects to CSL Behring's Pharmacovigilance Department at 1-866-915-6958.

*Based on April 2023 version.*



CSL Behring

RESEARCH

# ADVANCE-CIDP 3 Clinical Trial Results Favorable for Treating CIDP

Results of the Phase III ADVANCE-CIDP 3 clinical trial, a long-term extension study evaluating the safety and efficacy of HYQVIA (immune globulin infusion 10% [human] with recombinant human hyaluronidase) in patients with chronic inflammatory demyelinating polyneuropathy (CIDP) showed favorable long-term safety and tolerability of HYQVIA, and a low relapse rate, supporting its use as maintenance treatment for CIDP.

HYQVIA is the first and only facilitated subcutaneous immune globulin (fSCIG) approved by the U.S. Food and Drug Association as maintenance therapy in adults patients of all ages with CIDP post-stabilization with intravenous immune globulin (IVIG). HYQVIA's hyaluronidase component facilitates the dispersion and absorption of large IG volumes in the subcutaneous space between the skin and the muscle, allowing high-volume IG administration (equivalent to volumes administered intravenously) into the subcutaneous tissue over a short time. As a result, HYQVIA can be infused up to once monthly (every two, three or four weeks) and can be self-administered after appropriate training or administered by a healthcare professional in a medical office, infusion center or at a patient's home.

"The long-term data from the ADVANCE-CIDP 3 clinical trial allow us to further characterize the safety, efficacy and tolerability profile of HYQVIA and reinforces its role as a long-term, up to once monthly maintenance treatment for this complex, chronic condition," said Kristina Allikmets, senior vice president and head of research and development for Takeda's Plasma-Derived Therapies Business Unit. "These results reflect our continued commitment to bringing the benefits of our differentiated immune globulin therapies to patients with neuroimmunological disorders, and providing a range of effective treatment options that address the individual needs of a broad range of patients." IG

Takeda Presents Long-Term Data from Phase 3 ADVANCE-CIDP 3 Clinical Trial of HYQVIA in Patients with Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) at PNS Annual Meeting. BioSpace, June 18, 2024. Accessed at www.biospace.com/article/releases/takeda-presents-long-term-data-from-phase-3-advance-cidp-3-clinical-trial-of-hyqvia-in-patients-with-chronic-inflammatory-demyelinating-polyneuropathy-cidp-at-pns-annual-meeting.

RESEARCH

# CIDP Study Explores Dual-Target CAR-T Cell Therapy for the Treatment of Relapsed/Refractory Autoimmune Disorders

A team of researchers from the Cancer Institute of Xuzhou Medical University and the Affiliated Hospital of Xuzhou Medical University has revealed the feasibility, tolerability and efficacy of BCMA-CD19 bispecific CAR-T cells for treating chronic inflammatory demyelinating polyneuropathy (CIDP), highlighting the potential of CAR-T cell therapy in treating relapsed/refractory autoimmune disorders, which is a promising step toward creating a curable treatment for CIDP.

Treatments such as glucocorticoids, plasma exchange and intravenous immune globulin are currently used to help manage symptoms but cannot completely eradicate the disease. In recent years, CAR-T cell therapy has achieved remarkable efficacy in hematologic tumors and many diseases represented by systemic lupus erythematosus.

Researchers reported using BCMA-CD19 bispecific CAR-T cells for treating relapsed/refractory CIDP. Taking advantage of the BCMA protein's presence in plasma blasts and long-lived plasma cells, the researchers designed bispecific CAR-T cells that target both CD19 and BCMA, aiming to reset the balance of immune responses by temporarily and deeply eradicating B cells and plasma cells.

Recently, a patient diagnosed with distal CIDP without IgG4 autoantibodies made significant progress in functioning following CAR-T cell therapy, as per INCAT disability and MRC scores. Remarkably, almost full muscle power recovery was observed 180 days after CAR-T administration, paralleling the patient's ability to walk again. Post-treatment electrophysiological assessments of median, ulnar, common peroneal and tibial nerves also showed significant improvement. Remarkably, for more than a year, the patient could discontinue all immunosuppressants without disease recurrence, and the presence of GM4 and GD3 antibodies continued to diminish even after three months of CAR-T cell therapy.

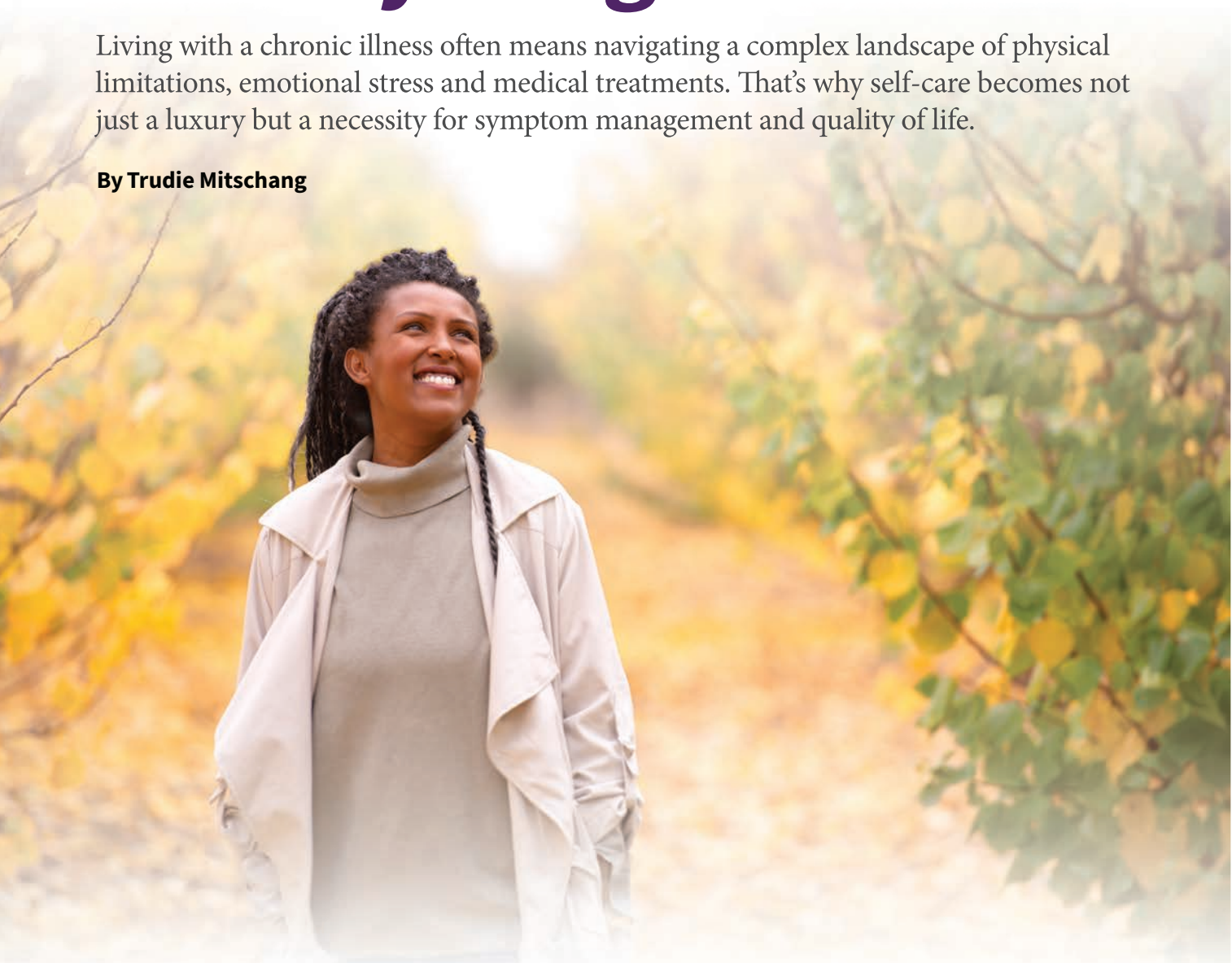
This study highlights the change in patient symptoms after treatment, affirms the safety of CAR-T cell therapy for CIDP and highlights the potential of CAR-T cell therapy in treating relapsed/refractory autoimmune disorders. IG

Tsinghua University Press. CIDP Study Explores Dual-Target CAR-T Cell Therapy for the Treatment of Relapsed/Refractory Autoimmune Disorders. Medical Xpress, June 20, 2024. Accessed at medicalxpress.com/news/2024-06-cidp-explores-dual-car-cell.html.

# Why Self-Care Is Anything But Selfish

Living with a chronic illness often means navigating a complex landscape of physical limitations, emotional stress and medical treatments. That's why self-care becomes not just a luxury but a necessity for symptom management and quality of life.

**By Trudie Mitschang**



**IN TODAY'S FAST**-paced world where a culture of busyness is often celebrated, the concept of self-care is often misunderstood and sometimes even stigmatized as being selfish. The truth is self-care is anything but self-centered, especially for those in the primary immune deficiency (PI) community who navigate myriad symptoms and struggles that their healthy counterparts cannot even fathom. When simply getting up and showered uses nearly all your daily "spoons," it's easy to see why self-care becomes a crucial component of disease management. When practiced correctly and regularly, it can help you function more effectively and live a more balanced and productive life — even with the limitations imposed by chronic illness.

"Self-care is hard when you struggle with depression, brain fog and fatigue. As much as I know things like exercise and meditation will help me feel better, it's sometimes easier said than done to include them in my routine," says patient advocate Eileen Davidson, who lives with rheumatoid arthritis (RA) and other chronic conditions.[1]

Alyssa Ebenezer, who was diagnosed with common variable immunodeficiency, agrees, noting that finding others in the PI community to talk with is a big stress-reliever that creates a support system for self-care. "I am a mom of two young kids, I work full time and deal with my chronic illness full time. I have learned to ask for help, and it has made such a difference. I've also connected with several women with the same diagnosis, and they are always available to talk. It really helps to have someone to speak with who 'gets it.'"[2]

### What Self-Care Is and Isn't

Instagram and TikTok stories typically promote self-care activities like candle-lit bubble baths or pricey spa days. While these activities can be helpful and stress-reducing, true self-care involves the body, mind and spirit, and provides the most benefits when incorporated into your daily lifestyle. According to an article published in the *British Medical Journal*, self-care is "the actions individuals take to lead a healthy lifestyle; to meet their social, emotional and psychological needs; to care for their long-term condition; and to prevent further illness or accidents."[3] In other words, it's about becoming proactive in caring for yourself rather than delegating that responsibility to loved ones and healthcare professionals.

The concept of self-care is not new. It actually originated in the 1950s, when doctors would recommend self-care as a way for institutionalized patients to exercise and treat themselves, often under medical supervision. It was later adopted by the civil rights movement of the 1960s as a mantra that promoted taking care of Black communities during times of great psychological stress. Later, self-care practices evolved to address the needs of those in high-risk professions and help them cope with the emotional impact of their jobs. Fast forward to 2016 when self-care became more of a main-stream buzzword thanks to social media, where it has been reinvented as a way that anyone can intentionally prioritize their own well-being.

While there is no one-size-fits-all method of self-care, most practices fit into one of seven basic pillars: mental, emotional, physical, environmental, spiritual, recreational and social. A well-balanced self-care routine involves each of these, and in practice, many will overlap. For example, physical and spiritual activities like taking a long walk or practicing meditation will have a positive impact on your mental and emotional state. Likewise, a recreational activity like going to hear live music is often a shared event, allowing you to check off the social pillar as well.

It's especially vital to practice self-care when you live with a chronic health condition that needs to be managed on a daily basis. Chronic diseases such as PI, autoimmune disorders and neurological conditions are often a major source of stress, which can lead to mental health conditions like anxiety and depression. Adapting self-care techniques such as mindfulness, daily journaling, meditation and deep breathing exercises can reduce stress levels and help you feel more in control of your condition. Keep in mind, self-care practices are very individualized and based on personal preferences and physical abilities and limitations. The important thing is to explore different practices and techniques to find the ones that feel most restorative for you.

## 5 Types of Self-Care

**Physical Self-Care:**
What we typically think of as self-care is caring for our physical body.

**Mental Self-Care:**
In the same way we work our muscles, we have to work our minds.

**Emotional Self-Care:**
We need to have systems for how we handle negative emotions such as journaling, therapy, etc.

**Spiritual Self-Care:**
We need to find connection with ourselves and others such as meditation, prayer, etc.

**Social Self-Care:**
Taking time for connection with others differs whether we're an extrovert or introvert.

* Adapted from: HyVee Health. What Is Self-Care and How Can You Practice It? Accessed at www.exemplar.care/news/what-is-self-care-and-how-can-you-practice-it.


## The S.T.O.P. Technique

**If you're feeling a moment of stress, try using the S.T.O.P Technique developed by mindfulness expert Jon Kabat-Zinn:**

**Stop:** Interrupt your thoughts and pause whatever you're doing. Sit up straight with a tall spine and calm yourself. Notice the sensations of being present.

**Take a breath:** Breathe in slowly through the nose, expanding the belly, and exhale slowly and deeply through pursed lips. Take a couple more conscious deep breaths, feeling the body relax with every inhale, and release tension with every exhale.

**Observe:** Become the observer of your thoughts, emotions and physical reactions. What thoughts do you notice? What emotions are present? How does your body feel? Tune in and sit with whatever arises for a few moments.

**Proceed:** Finally, proceed with intention and mindfully take each next step in your day from a place of strength, wisdom and presence.

### Self-Care and Stress Management

A 2022 Immune Deficiency Foundation (IDF) podcast titled "The Connection Between Mental and Physical Health" explored the challenges faced by those diagnosed with PI and offered specific ways to navigate those hurdles and improve the strength of both body and mind. Presented by John Seymore, PhD, LMFT, the episode explored how PI creates permanent stress in a person's life, including pain, discomfort and loss. "The diagnosis requires a lifetime of medical monitoring and sometimes is accompanied by additional health conditions — all of which generate medical expenses. PI brings unpredictability because, at one moment, a person may feel fatigued, and later, they regain their energy. Disruption occurs not only in a person's day but in the long term, causing uncertainty in starting a new job or school — and even in family planning," he explained.[4]

Dr. Seymore asserted that the best way to approach the seemingly overwhelming cycle of physical health leading to poor mental health is to reduce risk factors and increase protective factors: "Follow trusted sources of information pertaining to physical and mental health, create a self-care plan early on in diagnosis, and find trusted family members or friends who support you."

Practicing positivity whenever possible can also provide a boost to your mental health. Optimism has been shown to help frame the way your body reacts to a negative stressor; if you do not perceive an event to have a major impact on the state of your mood, then your body will not consciously perceive the stressor as that great of a threat. This means that an optimistic frame of mind can reduce the impact of a stress reaction, and the reduction of stress hormones can reduce inflammation in the body. This is where practicing an optimistic state of mind can lead to health benefits; your mindset can in itself be a type of self-care.

"When I was placed on long-term disability and stopped working as an aesthetician after my RA diagnosis, I struggled (and still struggle sometimes) with feeling like I don't have much to offer in this world. I felt like a burden," recalls Davidson. "But when I reflect on what I can still do, I can change that

negative inner dialogue around. I am a good mother who works hard to raise my son as best I can. I am an advocate, and my words help others cope. I volunteer. I create art. I take care of animals. I am a friend who people often come to for advice. This is so much healthier than focusing on my losses."[1]

Chronic disease blogger Alex Jean agrees: "On days you feel crummy, sometimes it's easy to get bogged down in those feelings. It can be easy to let those feelings consume you. At times, your chronic health problems can be a dominant voice you hear throughout the day. To help combat that voice when I'm having a flare day, I write down five things I'm grateful for that day. Some days are easier and some are tougher, but it does help to slightly shift the mindset."[5]

Alex Jean, who lives with three chronic illnesses, says journaling is also a go-to self-care habit she has developed to lower stress levels and suggests the following prompts for those new to the practice:

- How am I feeling today?
- What is contributing to how I am feeling today?
- How does my body feel today?
- How does my mind feel today?
- What is out of my control today?
- What is in my control today?
- What do I need today?

"Journaling doesn't have to be formal, or something you do every day. Journaling can also be on an as-needed basis when you feel you need to get things off your chest or organize your thoughts," she adds.

## Making Time for What Matters

While self-care can and often does involve prioritizing time for activities that feed your soul, when you live with a chronic illness, it also includes scheduling the things you need to get done that will help you feel your best, including:[6]

- *Medication management.* Plan your schedule so you take medications exactly as prescribed by your healthcare provider. Use pill organizers or apps to keep track.
- *Nutrition and meal planning.* The best way to ensure you consume a healthy balanced diet is to factor grocery shopping and meal prepping into your schedule. Not feeling up to a grocery store run? Consider an app like Instacart and get fresh fruits and veggies delivered — the small cost could well be worth the investment in better health.
- *Physical activity.* Performing some type of daily physical activity is essential for managing stress and inflammation levels. Gentle stretching, yoga and regular walks can all help improve mood and physical strength. Consider putting stretch breaks and daily walks on your calendar, and set reminders to stay on track.
- *Calendarize social activities.* Isolation is a real risk factor for those with chronic illness. Avoid this pitfall by putting social activities on your calendar. Even a weekly FaceTime with a best friend or loved one can dramatically boost your mood without draining your energy reserves.
- *Cultivate creativity.* Working in a garden. Drawing or finger painting. Writing poetry or short stories. These types of activities are known stress-reducers and offer health benefits to boot. Creative activity promotes something called "a state of flow," which has a restorative impact on our well-being and ultimately releases hormones that have a positive impact on our brains.

## Learning the Art of Self-Care

Learning the art of self-care is a dynamic, ongoing process that allows us to pay attention to our unique physical, mental, emotional and social needs. By incorporating customized self-care strategies into your daily routine, you can better manage your symptoms and potentially even foster a sense of empowerment and control over your treatment outcomes. The bottom line is you are not your illness, and prioritizing self-care allows you to focus on all of the aspects of what makes you unique.

"Self-love and self-care are a journey. It's normal to struggle, and I still often do. It's OK to not be OK, and it's definitely OK to reach out for help," says Davidson. "Research shows that those living with depression and anxiety tend to have poorer outcomes with arthritis or other chronic illness, so treating your mental health is as important as treating your physical health. But just because practicing self-love is hard doesn't mean it's not worth it. Without self-care and self-love, I wouldn't be me."[1] IG

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**TRUDIE MITSCHANG** is a contributing writer for *IG Living* magazine.

# The Spoon Theory, Reimagined?

The spoon theory provides an impactful explanation of how activities of daily living affect the energy levels of those living with chronic illness. Expanding upon its original concept can help enhance support and understanding for this community.

**By Surayyah Morris, PharmD**

**SPOON THEORY**, created in 2003 by Christine Miserandino who has lupus, offers a powerful metaphor that resonates deeply within the chronic illness community. It provides a way to articulate various unique struggles faced by those living with conditions such as immune deficiencies, chronic pain, lupus, cancer, fibromyalgia and a laundry list of other life-altering circumstances. With these types of ailments, it is imperative for you as a patient to treat yourself with grace, and invest your time in exercising the ability to manage your day-to-day activities as efficiently as possible, without triggering any negative symptoms or events. This blanket concept, which is the basis of spoon theory, is an important tool in the chronic warrior toolbox, and its versatility allows for reinventing efficiency in a way that is best for a positive quality of life.

Let's take a look at some factors that may influence a positive life experience for those of us who have a more difficult time than most. Not only will understanding these factors benefit you, the patient, but also your friends, family, doctors, politicians, teachers and caregivers — ultimately an entire community.

But first, let's understand spoon theory and how it works.

## What Is Spoon Theory?

Spoon theory is a way to understand the impact activities of daily living have on a person suffering from chronic illness or disability. This is significant because it highlights the impact of stress involved in simple functioning for those with chronic illness, compared to those who are able-bodied. It also validates how seemingly "manageable" tasks can be completely unbearable to complete.

Here's how spoon theory works: Picture yourself waking up each morning with a limited number of spoons. Each spoon represents a unit of energy needed to accomplish tasks throughout the day — simple tasks such as brushing teeth, getting dressed, cooking meals, walking or even socializing. For able-bodied healthy individuals, these activities might not consume much energy and are taken for granted. They

are performed with ease and without incident and are a basic part of a normal routine. However, for those with a chronic illness, each task requires careful consideration because it depletes their finite supply of spoons. Each movement is physically or mentally taxing. If you encounter others who tell you they're a spoonie, acknowledge this by respecting their limitations and boundaries, and always reserve patience for them.

### Applying the Spoon Theory

Having to budget energy is quite the dance, but a lively dance, nonetheless. Balancing tasks that are absolutely necessary during your day involves the knowledge of your physical and mental condition combined with adjustments to accommodate and facilitate meeting the goal.

1) *Daily energy budgeting.* Chronic illness warriors must prioritize tasks to conserve spoons for essential activities. This often means making tough choices. For instance, chores may have to be postponed to conserve energy for work or spending time with loved ones. Sometimes you must sacrifice things you want to do to save room for things you need to do or to simply circumvent an exacerbation of unwanted happenings.

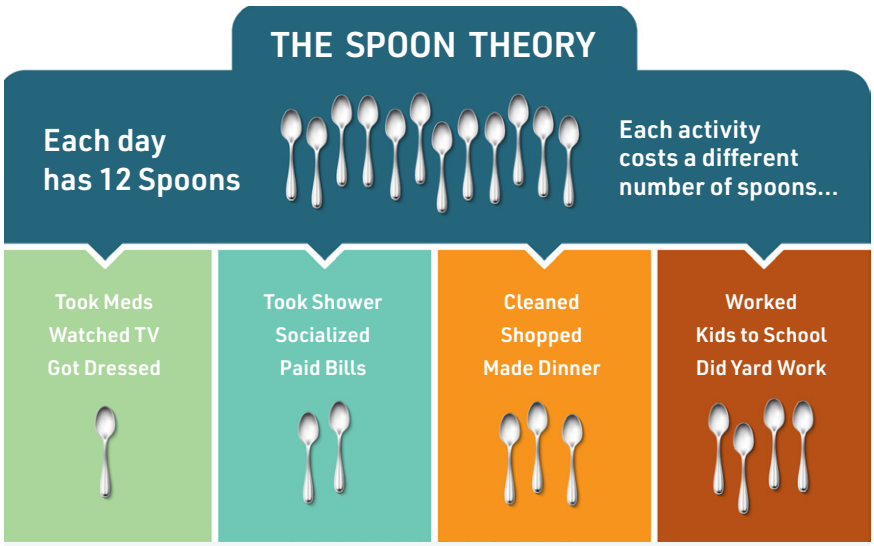
2) *Unpredictable nature.* The number of spoons available can fluctuate day to day or even hour to hour. Factors such as pain levels, medication side effects or stress can impact energy levels unpredictably, requiring constant adjustment and flexibility.

3) *Social and emotional impact.* The spoon theory also sheds light on the emotional toll of chronic illness. It can be frustrating when others don't understand why simple activities are exhausting or why plans change suddenly. This can lead to feelings of isolation or guilt for not being able to participate fully in daily life.

However, the spoon theory validates feelings of guilt or frustration that may arise from not being able to participate fully in activities or meet expectations. It also recognizes that limitations imposed by chronic illness builds self-compassion and provides encouragement to practice self-care without judgment.

### Communicating with Others

One of the most significant contributions of the spoon theory is its role in improving communication between chronic illness warriors and their loved ones. Using spoons




**A person has roughly the same amount of energy each day.**

- Each unit of energy is represented by a spoon.
- Healthy people have more spoons (energy) than those with an illness that causes chronic fatigue.
- Some activities cost more spoons than others.
- People with a fatiguing chronic illness need to make decisions about which activities to spend their limited number of spoons (energy) on – the illness stops them from doing many things they want to do.
- The chronic illness means that if people do too much in the morning, they will not have enough spoons left for the afternoon or evening.
- The chronic illness may mean people can do something in the morning or in the afternoon – but not both. They may seem inconsistent because their energy levels and number of spoons fluctuate.
- Healthy people do not need to plan how to spend their spoons because they do not have an illness that limits them.

gamunex®-C
immune globulin injection (human), 10%
caprylate/chromatography purified

# Step out of the symptoms of CIDP and back into your life with GAMUNEX-C

**GAMUNEX-C blocks the autoimmune and inflammatory processes of CIDP, which may improve symptoms and protect your nerves from damage.[1-3]***

*The way in which GAMUNEX-C works in treatment of people with CIDP is not completely understood.

GAMUNEX®-C (immune globulin injection [human], 10% caprylate/chromatography purified) is approved to treat primary humoral immunodeficiency disease (PIDD) in patients 2 years of age and older. If you have PIDD, you may take GAMUNEX-C under the skin (subcutaneously) or in a vein (intravenously). GAMUNEX-C is also approved to treat idiopathic thrombocytopenic purpura (ITP) in adults and children and chronic inflammatory demyelinating polyneuropathy (CIDP) in adults. If you have ITP or CIDP, you may only take GAMUNEX-C intravenously.

Do not take GAMUNEX-C if you have an allergy to immune globulin. Tell your doctor if you have had a serious reaction to other medicines that contain human immune globulin. Also tell your doctor if you have immunoglobulin A (IgA) deficiency. If you have a serious reaction while taking GAMUNEX-C, stop taking it immediately and tell your doctor.

CIDP, chronic inflammatory demyelinating polyneuropathy.

**Please see Important Safety Information for GAMUNEX-C on the following page.**

Learn more at GAMUNEX-C.com or call 1-888-MYGAMUNEX (1-888-694-2686)

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## IMPORTANT SAFETY INFORMATION

GAMUNEX®-C (immune globulin injection [human], 10% caprylate/chromatography purified) is approved to treat primary humoral immunodeficiency disease (PIDD) in patients 2 years of age and older. If you have PIDD, you may take GAMUNEX-C under the skin (subcutaneously) or in a vein (intravenously). GAMUNEX-C is also approved to treat idiopathic thrombocytopenic purpura (ITP) in adults and children and chronic inflammatory demyelinating polyneuropathy (CIDP) in adults. If you have ITP or CIDP, you may only take GAMUNEX-C intravenously.

**If you take GAMUNEX-C or a similar immune globulin product, you could experience a serious and life-threatening blood clot (thromboembolism), which may include pain and/or swelling of an arm or leg with warmth over the affected area, discoloration of an arm or leg, unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness, or weakness on one side of the body. You are more likely to develop a blood clot if you have a history of hardening of the arteries (atherosclerosis), stroke, heart attack, or heart failure (low volume of blood pumped by the heart). You may also be more likely to get a blood clot if you are elderly, if you have a blood clotting disorder, if you are inactive for long periods of time (such as long bed rest), if you use estrogens, or if you have thickening of your blood. For patients at risk, GAMUNEX-C should be administered at the lowest dose and slowest infusion rate that is practical. However, blood clots may occur in the absence of any of the known risk factors. Patients should be well hydrated by drinking enough water before GAMUNEX-C is administered. Tell your doctor immediately if your medical history is similar to what is described here, and especially if you start having any of these symptoms while taking GAMUNEX-C.**

**If you take GAMUNEX-C or a similar immune globulin product intravenously, you could experience serious kidney disease and death. You may have symptoms of decreased urination, sudden weight gain, swelling in your legs (edema), or shortness of breath. You are more likely to develop serious kidney disease if you already have a kidney problem, have Type II diabetes mellitus, or are older than 65. You are more likely to develop serious kidney disease if you are dehydrated, have a blood infection (sepsis), have high protein content in your blood, or if you are receiving other medicines that are harmful to your kidneys. Tell your doctor immediately if your medical history is similar to what is described here, and especially if you start having any of these symptoms while taking GAMUNEX-C.**

**You are more likely to develop serious kidney disease if you take an intravenous immune globulin product that contains sugar (sucrose). GAMUNEX-C does not contain sugar. If your situation makes you more likely to experience serious kidney disease, you should take GAMUNEX-C at the lowest concentration available and the slowest infusion rate that is practical.**

Do not take GAMUNEX-C if you have an allergy to immune globulin. Tell your doctor if you have had a serious reaction to other medicines that contain human immune globulin. Also tell your doctor if you have immunoglobulin A (IgA) deficiency. If you have a serious reaction while taking GAMUNEX-C, stop taking it immediately and tell your doctor.

Periodic monitoring of kidney function and urine output is particularly important in patients more likely to experience severe kidney disease.

You could experience other serious and life-threatening problems due to immune globulin. You could get aseptic meningitis (a type of brain inflammation with symptoms of severe headache, stiff neck, fatigue, fever, sensitivity to light, painful eye movements, nausea, and vomiting), a blood problem called hemolytic anemia (common symptoms include increased heart rate, fatigue, yellow skin or eyes, and dark-colored urine), and/or a lung problem called transfusion-related acute lung injury (commonly referred to as TRALI). TRALI is a condition where you build up fluid in the lungs (called pulmonary edema) that is not the result of heart failure.

If you have higher than normal body fluid volumes or if you have a condition where increasing body fluid volume may be a concern, a higher dose, such as 1g/kg for 1-2 days, is not recommended.

Because GAMUNEX-C is made from human blood, it may carry a risk of transmitting infectious agents such as viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent, and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.

You may not take GAMUNEX-C subcutaneously if you have ITP. **If you have ITP and take GAMUNEX-C subcutaneously, you could experience a very serious and life-threatening black and blue wound (hematoma, which is a pocket of blood within a tissue).**

After you take GAMUNEX-C, your blood antibody levels may rise, which could cause some blood antibody tests to give false results.

The most common side effects in a clinical study with PIDD patients who got subcutaneous injections of GAMUNEX-C were infusion-site reactions such as redness, swelling, and itching; extreme tiredness; pain in the region of the head or neck; a runny nose, nasal congestion, sneezing, cough, and sputum production; joint pain; loose stools; a sensation of unease and discomfort in the upper stomach; swelling of the tissue lining the sinuses; inflammation of the airways that carry air to your lungs; a feeling of unhappiness, sadness, melancholy, gloom, hopelessness, or low spirits; red rash or bumps, itchy, swollen, and tender skin with or without blisters or a burning feeling; a severe throbbing pain or a pulsing sensation, usually on just one side of the head; muscle pain; familiar infectious diseases such as the common cold or flu; and raised body temperature or fever. In clinical studies with PIDD patients who got GAMUNEX-C intravenously, the most common side effects were cough; irritation and inflammation of the mucous membrane inside the nose; sore throat caused by inflammation of the back of the throat; pain in the region of the head or neck; a condition in which your airways narrow and swell and produce extra mucus; a sensation of unease and discomfort in the upper stomach; raised body temperature or fever; loose stools; and swelling of the tissue lining the sinuses. In a clinical study with CIDP patients who got GAMUNEX-C intravenously, the most common side effects were pain in the region of the head or neck; raised body temperature or fever; abnormally high blood pressure; feelings of coldness accompanied by shivering; a noticeable change in the texture or color of your skin such as your skin becoming scaly, bumpy, itchy, or otherwise irritated; a sensation of unease and discomfort in the upper stomach; joint pain; and abnormal physical weakness or lack of energy. In clinical trials with ITP patients who got GAMUNEX-C intravenously, the most common side effects were pain in the region of the head or neck; a discoloration of the skin resulting from bleeding underneath, typically caused by bruising; vomiting, fever, nausea, rash, abdominal pain, back pain, and a pain or an uncomfortable feeling in the upper middle part of your stomach.

The most serious side effects in clinical studies were a blood clot to the lung (pulmonary embolism) in 1 patient with a history of this condition (in CIDP), a flare-up of an existing type of anemia (autoimmune pure red cell aplasia) in 1 patient (in PIDD), and heart inflammation (myocarditis) in 1 patient (in ITP).

**Please see brief summary of the full Prescribing Information on the following page.**

**You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch or call 1-800-FDA-1088.**

**References: 1.** Merkies IS, Bril V, Dalakas MC, et al. Health-related quality-of-life improvements in CIDP with immune globulin IV 10%: the ICE Study. *Neurology.* 2009;72(15):1337-1344. **2.** Hughes RAC, Donofrio P, Bril V, et al; on behalf of the ICE Study Group. Intravenous immune globulin (10% caprylate-chromatography purified) for the treatment of chronic inflammatory demyelinating polyradiculoneuropathy (ICE study): a randomised placebo-controlled trial. *Lancet Neurol.* 2008;7(2):136-144. **3.** Dalakas MC, Latov N, Kuitwaard K. Intravenous immunoglobulin in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): mechanisms of action and clinical and genetic considerations. *Expert Rev Neurother.* 2022;22(11-12):953-962.

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# GAMUNEX®-C

## Immune Globulin Injection (Human), 10% Caprylate/Chromatography Purified

### HIGHLIGHTS OF PRESCRIBING INFORMATION

**These highlights do not include all the information needed to use GAMUNEX®-C safely and effectively. See full prescribing information for GAMUNEX-C.**

**GAMUNEX®-C, [Immune Globulin Injection (Human), 10% Caprylate/Chromatography Purified]**

**Initial U.S. Approval: 2003**

**WARNING: THROMBOSIS, RENAL DYSFUNCTION and ACUTE RENAL FAILURE**

***See full prescribing information for complete boxed warning.***

- **Thrombosis may occur with immune globulin products, including GAMUNEX-C. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **For patients at risk of thrombosis, administer GAMUNEX-C at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**
- **Renal dysfunction, acute renal failure, osmotic nephrosis, and death may occur with immune globulin intravenous (IGIV) products in predisposed patients.**
- **Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. GAMUNEX-C does not contain sucrose.**
- **For patients at risk of renal dysfunction or failure, administer GAMUNEX-C at the minimum concentration available and the minimum infusion rate practicable.**

### ---------- INDICATIONS AND USAGE ----------

GAMUNEX-C is an immune globulin injection (human), 10% liquid indicated for treatment of:

- Primary Humoral Immunodeficiency (PI) in patients 2 years of age and older
- Idiopathic Thrombocytopenic Purpura (ITP) in adults and children
- Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) in adults

### ---------- DOSAGE AND ADMINISTRATION ----------

**Intravenous Administration Only: ITP and CIDP**

| Indication | Dose | Initial Infusion Rate | Maintenance Infusion Rate (if tolerated) |
|---|---|---|---|
| ITP | 2 g/kg | 1 mg/kg/min | 8 mg/kg/min |
| CIDP | loading dose 2 g/kg<br>maintenance dose 1 g/kg | 2 mg/kg/min | 8 mg/kg/min<br>Every 3 weeks |

- Ensure that patients with pre-existing renal insufficiency are not volume depleted; discontinue GAMUNEX-C if renal function deteriorates.
- For patients at risk of renal dysfunction or thrombosis, administer GAMUNEX-C at the minimum infusion rate practicable.

**Intravenous or Subcutaneous Administration: PI**
**DO NOT ADMINISTER SUBCUTANEOUSLY FOR ITP PATIENTS**

| Route of Administration | Dose | Initial Infusion Rate | Maintenance Infusion Rate (if tolerated) |
|---|---|---|---|
| Intravenous (IV) | 300-600 mg/kg | 1 mg/kg/min | 8 mg/kg/min<br>Every 3 to 4 weeks |
| Subcutaneous (SC) | 1.37 x current IV dose in grams/ IV dose interval in weeks | Adult:†<br>20 mL/hr/site<br>Pediatric:†<br>10 mL/hr/site (< 25 kg)<br>15 mL/hr/site (≥ 25 kg) | Adult:†<br>20 mL/hr/site<br>Pediatric:†<br>10 mL/hr/site (< 25 kg)<br>20 mL/hr/site (≥ 25 kg) Weekly |

† Adults: use up to 8 infusion sites simultaneously; pediatric: use up to 6 infusion sites simultaneously; for all ages, ensure infusion sites are at least 2 inches (5 cm) apart.

### ---------- DOSAGE FORMS AND STRENGTHS ----------

GAMUNEX-C is a sterile solution for injection supplied in 1 g (10 mL), 2.5 g (25 mL), 5 g (50 mL), 10 g (100 mL), 20 g (200 mL), or 40 g (400 mL) single use vials.

### ---------- CONTRAINDICATIONS ----------

- Anaphylactic or severe systemic reactions to human immunoglobulin
- IgA deficient patients with antibodies against IgA and a history of hypersensitivity

### ---------- WARNINGS AND PRECAUTIONS ----------

- IgA deficient patients with antibodies against IgA are at greater risk of developing severe hypersensitivity and anaphylactic reactions. Have epinephrine available immediately to treat any acute severe hypersensitivity reactions.
- Hyperproteinemia, with resultant changes in serum viscosity and electrolyte imbalances may occur in patients receiving IGIV therapy.
- Aseptic Meningitis Syndrome (AMS) may occur, especially with high doses or rapid infusion.
- Hemolysis, either intravascular or due to enhanced RBC sequestration, can develop subsequent to GAMUNEX-C treatments. Risk factors include high doses and non-O blood group. Closely monitor patients for hemolysis and hemolytic anemia, especially in patients with pre-existing anemia and/or cardiovascular or pulmonary compromise.
- Monitor patients for pulmonary adverse reactions (transfusion-related acute lung injury [TRALI]).
- Volume overload.
- GAMUNEX-C is made from human plasma and may carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent.
- GAMUNEX-C is not approved for subcutaneous use in ITP patients. Due to a potential risk of hematoma formation, do not administer GAMUNEX-C subcutaneously in patients with ITP.
- Passive transfer of antibodies may confound serologic testing.

### ---------- ADVERSE REACTIONS ----------

The most common adverse reactions observed in ≥ 5% patients were:

**PI**: Intravenous: Cough increased, rhinitis, pharyngitis, headache, asthma, nausea, fever, diarrhea, and sinusitis.
Subcutaneous: Local infusion site reactions, fatigue, headache, upper respiratory tract infection, arthralgia, diarrhea, nausea, sinusitis, bronchitis, depression, allergic dermatitis, erythema, migraine, myalgia, viral infection, and pyrexia.

**ITP**: Headache, ecchymosis, vomiting, fever, nausea, rash, abdominal pain, back pain, and dyspepsia.

**CIDP**: Headache, pyrexia, hypertension, chills, rash, nausea, arthralgia, and asthenia.

**To report SUSPECTED ADVERSE REACTIONS, contact Grifols Therapeutics LLC at 1-800-520-2807 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.**

### ---------- DRUG INTERACTIONS ----------

- The passive transfer of antibodies may transiently interfere with the response to live virus vaccines, such as measles, mumps and rubella.

### ---------- USE IN SPECIFIC POPULATIONS ----------

- Geriatric: In patients over 65 years of age do not exceed the recommended dose, and infuse GAMUNEX-C at the minimum infusion rate practicable.

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**3054846/3054847**
**Revised: 1/2020**

as a metaphor, you can effectively convey the concept of limited energy reserves by:

1) *Educating others.* Sharing the spoon theory helps friends, family and coworkers understand the daily challenges faced by those with chronic illnesses. It fosters empathy and encourages a supportive environment.

2) *Setting boundaries.* Knowing your limits allows you to set boundaries without feeling guilty. Saying "no" to nonessential activities becomes a necessary act of self-care rather than a reflection of unwillingness or disinterest.

### Embracing the Spoon Theory as Empowerment

While the spoon theory acknowledges the difficulties of living with chronic illness, it also emphasizes empowerment through:

• *Advocacy:* It encourages advocacy for better understanding and accommodations in various aspects of life, including workplaces, social settings and healthcare.

• *Building support networks:* The spoon theory creates a sense of community among individuals facing similar challenges, providing mutual support and understanding. This is also extremely important for validation of experiences.

**As our understanding of chronic illness evolves and new perspectives emerge, there's an opportunity to explore innovative approaches that complement and expand upon the original spoon theory concept.**

• *Self-acceptance:* Accepting the limitations imposed by illness is a crucial step toward self-compassion and resilience.

• *Adaptability:* Learning to adapt daily routines and expectations helps you make the most of the spoons you have available.

As a cornerstone in the chronic illness community, the spoon theory offers a vocabulary to articulate experiences that are often misunderstood or overlooked. It provides a framework for self-care and advocacy, and it builds understanding among peers and loved ones. By embracing the spoon theory, you can navigate your journey with greater resilience, dignity and support.

In essence, the spoon theory reminds us all to cherish the energy we have, to support those with chronic illnesses with empathy and to recognize the strength and courage it takes to live each day to the fullest despite the challenges faced.

### Reinventing the Spoon Theory: Innovating Support for Chronic Conditions

As our understanding of chronic illness evolves and new perspectives emerge, there's an opportunity to explore innovative approaches that complement and expand upon the original spoon theory concept. Let's consider a few things:

**Redefining Energy and Resources**

1) *Expanded definition of energy.* Beyond physical and mental energy, a reinvented theory can encompass a broader spectrum of resources. This might include emotional resilience, cognitive bandwidth and the energy expended in navigating societal barriers and stigma.

2) *Intersectional approach.* Recognizing how disability interconnects with other identities (such as race, gender, sexuality and socioeconomic status) is crucial. An updated framework could address how these intersections impact energy management and access to resources. Involving the spoonie community and merging with the established organizations can unify so many more, producing more information, data and experiences that lead to a variety of management strategies when dealing with chronic illness.

**Accessibility and Inclusivity**

1) *Universal design.* Incorporating principles of universal design into the reinvented theory can ensure it accommodates diverse disabilities and communication styles. This includes making resources available in multiple formats (e.g., visual, auditory, tactile) and ensuring accessibility in digital and physical environments. In this day and age, this would seem like an unnecessary suggestion; however, there is still a barrier to the availability of accessible resources.

2) *Cultural sensitivity.* Acknowledging cultural differences and varying perspectives on disability is essential. The

reinvented spoon theory should promote inclusivity and respect for diverse experiences within the disabled community.

**Empowerment and Advocacy**

1) *Empowering self-advocacy.* Providing tools and strategies for self-advocacy is crucial. This includes promoting assertiveness in communicating needs, navigating systems and advocating for accessible environments. This would empower individuals to share insights, strategies and resources for managing chronic conditions.

2) *Community support.* Strengthening community support networks and peer mentoring can enhance resilience and solidarity within the disabled community. Platforms for sharing experiences, resources and strategies can facilitate mutual aid and empowerment. An updated theory could emphasize education about diverse chronic conditions, their varying impacts and the importance of empathy and support. This would aim to reduce stigma and generate a more inclusive understanding of chronic illness.

**Practical Applications and Innovation**

1) *Technology and accessibility tools and apps.* Leveraging technology can enhance accessibility and communication, both pivotal to supporting use of the spoon theory. This might involve digital trackers, activity planners, apps for energy management, adaptive technologies and assistive devices that support independence and participation in daily life. Remember: In this day and age, "there's an app for that." If there isn't an app currently, there soon will be, so don't worry. These innovations assist in communicating needs effectively to caregivers and healthcare providers.

2) *Personalized metrics.* Recognizing that each individual's experience of chronic illness is unique, a reinvented spoon theory could encourage the development of personalized metrics. This might involve quantifying energy reserves based on specific symptoms, medication impacts or lifestyle factors. Embracing the knowledge that technology provides in assisting with the mind and body as specifically as possible is monumental in reimagining how you use the spoon theory to manage day to day.

3) *Policy and structural change.* Advocating for policy reforms and structural changes is vital for creating inclusive environments. This includes promoting accessible transportation, housing, employment opportunities and healthcare services. These services are available and are plentiful in some areas, but may still require a few additional spoons to navigate locating and obtaining services that will benefit your situation. These spoons are OK to use though; consider it an investment in a comfortable future.

4) *Evolving perspectives on defining energy and health fluidity of energy.* While the spoon theory illustrates energy as a finite resource, there's recognition that energy levels can fluctuate dynamically. Introducing a concept of "energy waves" or "energy reservoirs" could better capture the ebb and flow experienced by individuals with chronic conditions. This acknowledges that some days might offer more reserves, while others require careful conservation.

5) *Holistic wellness.* Beyond physical energy, chronic conditions impact emotional, cognitive and social aspects of life. A reinvented theory could integrate these dimensions, emphasizing the interconnectedness of mental health, social support and overall well-being.

**Practical Applications in Daily Life**

1) *Flexible strategies.* Encouraging flexibility in energy management strategies can empower you to adapt to changing circumstances. This might involve promoting mindfulness, pacing techniques and adaptive planning.

2) *Integration with healthcare.* Reinventing the spoon theory could promote its integration into healthcare settings. Healthcare providers may find use in updating frameworks to better assess patient needs, tailor treatment plans and support patient education. If your providers are open and willing, it may benefit you to introduce them to your experience to facilitate change for yourself and possibly for other patients as well.

## Reinventing the Spoon Theory: An Opportunity

Reinventing the spoon theory presents an opportunity to reimagine and enhance support and understanding for individuals living with chronic conditions. By embracing evolving perspectives on energy management, leveraging technology for communication and empowerment, and promoting holistic wellness, we can build upon the foundation laid by the spoon theory. Ultimately, these innovations aim to empower individuals, foster community resilience and advance compassionate care for all those navigating the challenges of chronic illness. IG

**SURAYYAH MORRIS**, PharmD, is an IG patient from Central Florida. As a medication therapy management and pain management specialty pharmacist, she enjoys supporting patients with chronic pain and chronic conditions to help find balance and improve quality of life.

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# 7 Foods for a Better Night's Sleep

From sweet to soothing, these bedtime snacks may be just what you need for a more restful sleep.

**By Emily Cooper, RDN**

**IT MAY BE** well-known that eating nutritiously and staying active are keys to good health, but did you know getting enough sleep is just as important? More than one-third of American adults get less than seven hours of sleep a night. Not getting enough sleep can put you at greater risk for diabetes, obesity and heart disease. A lack of sleep can also increase irritability and decrease concentration and overall brain performance. The recommendation for most American adults is to aim for seven to nine hours of quality sleep every night.[1] Even one night of inadequate sleep can impact your overall physical and mental well-being.

Certain things such as a cool, dark room and a nighttime routine can set you up for sleep success, but what you eat before bed can also impact your restfulness. Here are seven foods to add to your diet, and a few to avoid when it comes to getting quality shuteye.

### Kiwi

If you like to end your night meal on a sweet note, enjoying some fresh kiwi can both satisfy your sweet tooth and help you sleep better. Kiwi contains the sleep hormone melatonin, which helps regulate the body's internal clock, also known as circadian rhythm. Melatonin also helps calm the body to prepare for sleep. Research has shown that eating kiwi before bed can help you fall asleep faster, improve sleep quality and help you sleep longer. Kiwi is also a natural source of antioxidants such as vitamin C, which have also been shown to be associated with better-quality sleep.[2]

You can enjoy fresh kiwi as is, blend into a smoothie or chop into a salsa with fresh cilantro, lime juice and bell peppers. Most research has shown the best benefits with a serving of one to two kiwis per night.

### Nuts

If crunchy is more your style, a handful of dry roasted nuts such as almonds, walnuts or cashews are the perfect sleep-promoting snack. Nuts are a natural source of magnesium and calcium. Higher levels of magnesium are associated with better and longer sleep and falling asleep faster.[3] Getting enough calcium is associated with ease of falling asleep and more restorative sleep.

Nuts are also a natural source of unsaturated fats and protein. The healthy fats in nuts can help with serotonin levels, preparing the body for a restful night's sleep. Along with the protein in nuts, these healthy fats help to keep blood sugar levels stable throughout the night. Stable blood sugars also help to promote restful sleep.

Opt for dry-roasted nuts without added salt or sugars. Enjoy a handful of mixed nuts as a nighttime snack, sprinkle over a salad at dinner, or chop and add to a bowl of fresh fruit for an added crunch.

### Dairy

A glass of warm milk before bed may be more than old folklore. Dairy foods such as milk, yogurt and cheese are calcium-rich. This bone-building nutrient is also needed to produce melatonin, which helps prepare the body for sleep. Dairy foods also contain tryptophan, an amino acid that has been associated with better sleep and overall mood. Tryptophan also helps with the production of melatonin and serotonin in the body.[4]

Some ideas for including dairy in your diet before bed (aside from a glass of milk) are enjoying a bowl of plain Greek yogurt topped with fresh fruit or chopped nuts, pairing a serving of your favorite cheese with a piece of fruit, or a bowl of low-sodium cottage cheese sprinkled with fresh herbs.

### Tart Cherries

If a glass of warm milk isn't your thing, tart cherry juice may be a welcome alternative. Cherries, especially tart cherries, are a natural source of melatonin and tryptophan, both of which contribute to restfulness and quality sleep. The tart cherry variety such as Montmorency contains much higher amounts of melatonin than sweeter varieties like Bing or Rainier.[5] Montmorency cherries are much more sour and tart, and a lot of times are consumed more in the form of juice than eating as is.

Cherries overall contain antioxidants like vitamins A and C, which have been shown to play a positive role in sleep quality. If you opt for a cherry juice, make sure it is 100 percent tart cherry juice with no added sugars. If it is too tart to drink on its own, mix it with sparkling or plain water. Be sure to talk to your primary care provider, especially if you are on any blood-thinning medications, before starting to drink tart cherry juice. You can still get some of the sleep-promoting benefits from regular cherries. Enjoy them fresh, blend frozen cherries into a smoothie or use as a topping over Greek yogurt.

### Fish

If you're looking for a sleep-promoting dinner option, looking to the sea may be your answer. Regularly consuming fatty fish, which includes salmon, mackerel, sardines, anchovies and herring, is associated with better-quality sleep. Fish is also a source of sleep-promoting nutrients such as vitamin D, omega-3 fatty acids, vitamin B6 and tryptophan. Tryptophan and vitamin B6 are nutrients required for the body to produce melatonin. Omega-3 fatty acids have been associated with healthy sleep duration and overall sleep quality.[6]

The U.S. Department of Agriculture recommendation for fish and seafood consumption for American adults is about eight ounces, or roughly two servings per week. While fatty fish contain the highest levels of omega-3 fatty acids, other choices to consider include cod, halibut, tuna and snapper.

### Bananas

Another choice for the sweet lovers, bananas are a sleep-friendly food to enjoy before bed. They are a natural source of both potassium and magnesium, which are nutrients that


## 7 Foods Containing Ingredients That Boost Sleep

support muscle relaxation. Bananas also contain tryptophan, which has been associated with quality sleep and can positively impact mood. This popular fruit also contains antioxidants such as vitamin C, which supports quality sleep and overall health.

For extra sleep-inducing effects, pair your banana with a handful of mixed nuts, a bowl of Greek yogurt or mix with chopped up kiwi and cherries for a sleepy bowl of fruit salad.

### Tea

Is there anything more comforting at the end of the day than a cup of hot tea? Aside from warming the body and soul, a cup of tea can also positively impact sleep. Herbal teas, especially peppermint and chamomile, are most often associated with promoting sleepiness. Chamomile contains apigenin, a flavonoid that is known for its soothing and calming effects.[7] Peppermint is most often associated with reducing stress and anxiety and promoting sleep, both of which make it a great choice for a nighttime beverage.

If peppermint or chamomile teas aren't your favorite, enjoy a cup of your favorite tea instead. Including this step at the end of each day can foster a relaxing routine, preparing your body and mind for sleep. Be sure to opt for a caffeine-free tea to avoid any sleep disruptions.

### What to Avoid

Just as there are foods that can help promote healthy sleep, there are also certain things that can hinder it. Here are some things to avoid right before bedtime to prevent sleep disturbances.

*Caffeine.* While herbal teas can do wonders for extra sleep, caffeinated teas, coffee and other beverages can make falling asleep harder and interrupt sleep patterns. Aside from these drinks, another lesser-known source of caffeine is dark chocolate. A one-ounce serving of dark chocolate contains about 24 mg of caffeine, which is roughly a quarter the amount of caffeine as a cup of coffee. While it may not seem like a significant amount of caffeine, it can impact sleep for those who are sensitive to caffeine's effects, especially if multiple servings of dark chocolate are eaten at once.

*Alcohol.* Enjoying a nightcap at the end of the day can help some relax, unwind and fall asleep easier, but it can actually disrupt restful sleep overall. Having alcohol in your bloodstream when you go to sleep can impact your regular sleep cycle, leading to poorer sleep quality and lower amounts of total sleep. A moderate serving of alcohol, which is two drinks for men and one drink for women, can decrease overall sleep quality by up to 24 percent. Alcohol can also make sleep disorders such as insomnia or sleep apnea worse.[8]

If you want to enjoy a drink at the end of the day, give your body enough time to process it before going to bed to prevent sleep disruptions. Alternatively, enjoy a cup of herbal tea as a way to wind down without sacrificing quality shuteye.

*Spicy or heavy foods.* Have you ever noticed how spicy foods can seem to wake you up? That may be something you want to avoid when trying to get to sleep. Spicy foods can also increase your body temperature and induce heartburn, making restful sleep more difficult.

Heavy foods such as cream sauces, large steaks, fried foods or dishes with a lot of cheese can take longer for the body to digest. This can lead to indigestion, making it harder to get comfortable and fall asleep at night. If you want to enjoy these types of foods, give your body enough time to properly digest them before going to bed. Usually two to four hours is enough time between your last meal and sleep. Keep an eye out for any foods you find particularly bother you or lead to sleep disruptions, and avoid them before bed.

### Sleep Is Important for Overall Health

Getting the sleep your body needs to rest and restore is important for overall health. While having an overall balanced diet, moving your body and managing stress may be highly important for fostering an environment for rest, adding or avoiding certain foods can help give you the boost you need to wake up ready to take on the day. IG

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**If you're an adult living with primary immunodeficiency (PI), ALYGLO™ can reduce the risk of infection from PI and its impact on your daily life.**[1]
Based on a clinical study of 33 adults ages 17-70 in North America.[1]

**0.03** SERIOUS INFECTIONS per patient year[1]

**0.2** DAYS OF HOSPITALIZATION per patient year[1]

**6** DAYS MISSED OF WORK OR SCHOOL per year[1]

## INDICATION

ALYGLO™ is indicated for the treatment of primary humoral immunodeficiency (PI) in adults aged 17 years and older. This includes, but is not limited to, congenital agammaglobulinemia, common variable immunodeficiency (CVID), Wiskott-Aldrich syndrome, and severe combined immunodeficiencies.

## IMPORTANT SAFETY INFORMATION

- **Thrombosis (blood clot formation) can happen with ALYGLO. Factors that increase this risk include advanced age, prolonged immobility, certain medical conditions, and cardiovascular risk factors.**
- **ALYGLO may affect the kidneys. In some cases, it can lead to acute renal failure or death.**
- **If you're at risk for blood clots or kidney problems, your doctor should give you ALYGLO at the lowest effective dose and infusion rate. Staying well-hydrated before treatment is essential.**
- ALYGLO is not suitable for people who have had severe allergic reactions to immune globulin or those with IgA deficiency and a history of hypersensitivity.
- If you experience any signs of hypersensitivity during the infusion, treatment should be stopped and epinephrine (an emergency medication) should be administered immediately.
- ALYGLO may cause hyperproteinemia, increased serum viscosity, and hyponatremia (low sodium levels).
- Aseptic Meningitis Syndrome (AMS) is a rare condition that can occur after receiving ALYGLO, especially with high doses or rapid infusion. Symptoms usually start within a few hours to 2 days after treatment. If AMS occurs, stopping ALYGLO usually leads to improvement within several days without lasting effects.
- Hemolysis, a breakdown of red blood cells, may occur. Some patients may experience delayed hemolytic anemia due to increased sequestration of red blood cells. Severe hemolysis-related kidney dysfunction or disseminated intravascular coagulation has been reported.
- Transfusion-Related Acute Lung Injury (TRALI) is a rare complication characterized by severe respiratory distress, pulmonary edema, and fever. Patients with TRALI may need oxygen therapy and ventilator support.
- ALYGLO is made from human blood, which may carry a risk of transmitting infectious agents (such as viruses).
- After receiving ALYGLO, some antibodies from the treatment may temporarily show up in blood tests. This could lead to misleading results, so your healthcare provider will consider this when interpreting lab results.
- Common side effects include headache, nausea/vomiting, fatigue, nasal/sinus congestion, rash, arthralgia, diarrhea, muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, and dizziness.

**Reference: 1.** ALYGLO Prescribing Information. GC Biopharma; 2023.

**For more information about ALYGLO, talk to your doctor and see Brief Summary of Prescribing Information on next page.**



GC Biopharma

**Alyglo™**
*immune globulin intravenous, human-stwk 10% liquid*

# BRIEF SUMMARY OF PRESCRIBING INFORMATION
## Please see full Prescribing Information at ALYGLO.com.

**WARNING: THROMBOSIS, RENAL DYSFUNCTION and ACUTE RENAL FAILURE**

***See full prescribing information for complete boxed warning.***

- **Thrombosis may occur with immune globulin intravenous (IGIV) products, including ALYGLO. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **Renal dysfunction, acute renal failure, osmotic nephropathy, and death may occur with the administration of IGIV products in predisposed patients.**
- **Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. ALYGLO does not contain sucrose.**
- **For patients at risk of thrombosis, renal dysfunction or renal failure, administer ALYGLO at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**

## INDICATIONS AND USAGE

ALYGLO is a 10% immune globulin liquid for intravenous injection, indicated for the treatment of primary humoral immunodeficiency (PI) in adults. This includes, but is not limited to, the humoral immune defect in congenital agammaglobulinemia, common variable immunodeficiency (CVID), X-linked agammaglobulinemia, Wiskot-Aldrich syndrome, and severe combined immunodeficiency (SCID).

## DOSAGE AND ADMINISTRATION

**For intravenous use only.**

**Dose**

**Table 1 Recommended Dose**

| Dose | Infusion Number | Initial Infusion Rate | Maintenance Infusion Rate |
|---|---|---|---|
| 300 - 800 mg/kg body weight every 21 or 28 days | For the 1st Infusion | 1 mg/kg/min (0.01 mL/kg/min) | Double the infusion rate every 30 minutes (if tolerated) up to 8 mg/kg/min (0.08 mL/kg/min) |
| 300 - 800 mg/kg body weight every 21 or 28 days | From the 2nd Infusion | 2 mg/kg/min (0.02 mL/kg/min) | Double the infusion rate every 15 minutes (if tolerated) up to 8 mg/kg/min (0.08 mL/kg/min) |

Significant differences in the half-life of IgG among patients with PI may necessitate the dose and frequency of immunoglobulin therapy to vary from patient to patient. Determine the proper dose and frequency by monitoring clinical response.

Measles Exposure

If a patient has been exposed to measles, consult with physician to administer an extra dose of IGIV as soon as possible and within 6 days of exposure. A dose of 400 mg/kg should provide a serum level > 240 mIU/mL of measles antibodies for at least two weeks.

If a patient is at risk of future measles exposure and receives a dose of less than 530 mg/kg every 3 - 4 weeks, then the dose should be increased to at least 530 mg/kg. This should provide a serum level of 240 mIU/mL of measles antibodies for at least 22 days after infusion.

**Administration**

- Monitor vital signs throughout the infusion. Slow or stop the infusion if adverse reactions occur. If symptoms subside, the infusion may be resumed at a lower rate that is comfortable for the patient.
- Ensure that patients with pre-existing renal insufficiency are not volume depleted. For patients at increased risk of renal dysfunction or thrombotic events, administer ALYGLO at the minimum infusion rate practicable, and consider discontinuation of administration if renal function deteriorates [*see Boxed Warning, Warnings and Precautions*].
- After administration, the infusion line may be flushed with either normal saline or 5% dextrose in water.

## CONTRAINDICATIONS

ALYGLO is contraindicated in:

- Patients who have a history of anaphylactic or severe system reaction to the administration of human immune globulin.
- IgA-deficient patients with antibodies to IgA and a history of hypersensitivity [*see Warnings and Precautions*].

## WARNINGS AND PRECAUTIONS

**Hypersensitivity:** Severe hypersensitivity reactions may occur[1]. In case of hypersensitivity, discontinue ALYGLO infusion immediately and institute appropriate treatment. Have epinephrine available for immediate treatment of severe acute hypersensitivity reactions.

ALYGLO contains trace amounts of IgA (≤ 100 mcg/mL). Patients with known antibodies to IgA may have a greater risk of developing potentially severe hypersensitivity and anaphylactic reactions. ALYGLO is contraindicated in IgA-deficient patients with antibodies against IgA or a history of hypersensitivity reaction [*see Contraindications*].

**Thrombotic Events:** Thrombosis may occur following treatment with ALYGLO[1]. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling central vascular catheters, hyperviscosity and cardiovascular risk factors. Thrombosis may occur in the absence of known risk factors.

Consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including patients with cryoglobulins, fasting chylomicronemia/markedly high triacylglycerols (triglycerides), or monoclonal gammopathies. For patients at risk of thrombosis, administer ALYGLO at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity [*see Boxed Warning, Dosage and Administration*].

**Renal Failure:** Renal dysfunction, acute renal failure, osmotic nephropathy, and death[1] may occur upon use of ALYGLO. Ensure that patients are not volume-depleted before administering ALYGLO. Monitor renal function and urine output periodically, especially in patients who are at higher risk of renal failure. Assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine before the initial infusion of ALYGLO and at appropriate intervals thereafter. If renal function deteriorates, consider discontinuing ALYGLO. In patients who are at risk of developing renal dysfunction, because of pre-existing renal insufficiency or predisposition to acute renal failure (such as diabetes mellitus, hypovolemia, overweight, use of concomitant nephrotoxic medicinal products or age > 65 years), administer ALYGLO at the minimum infusion rate practicable [*see Boxed Warning, Dosage and Administration*].

**Hyperproteinemia, Increased Serum Viscosity, and Hyponatremia:** Hyperproteinemia, increased serum viscosity, and hyponatremia may occur in patients receiving ALYGLO. It is critical to clinically distinguish true hyponatremia from a pseudohyponatremia that is associated with or causally related to hyperproteinemia with concomitant decreased calculated serum osmolality or elevated osmolar gap. Such treatment aimed at decreasing serum free water in patients with pseudohyponatremia may lead to volume depletion, a further increase in serum viscosity, and a possible predisposition to thrombotic events[1].

**Aseptic Meningitis Syndrome (AMS):** AMS may occur with ALYGLO. AMS usually begins within several hours to 2 days following ALYGLO treatment. Discontinuation of treatment has resulted in remission of AMS within several days without sequelae[1].

AMS may occur more frequently with high doses (2 g/kg) and/or rapid infusion of ALYGLO. AMS is characterized by the following signs and symptoms: Severe headache, nuchal rigidity, drowsiness, fever, photophobia, painful eye movements, nausea, and vomiting. Cerebrospinal fluid (CSF) studies frequently reveal pleocytosis up to several thousand cells per cubic millimeter, predominantly from the granulocytic series, and elevated protein levels up to several hundred mg/dL, but negative culture results. Conduct a thorough neurological examination on patients exhibiting such signs and symptoms, including CSF studies, to rule out other causes of meningitis.

**Hemolysis:** ALYGLO may contain blood group antibodies that can act as hemolysins and induce *in vivo* coating of red blood cells (RBCs) with immunoglobulin, causing a positive direct antiglobulin test (DAT) (Coombs test) result and hemolysis[1]. Delayed hemolytic anemia due to enhanced RBC sequestration, and acute hemolysis, consistent with intravascular hemolysis, have been reported. Cases of severe hemolysis-related renal dysfunction/failure or disseminated intravascular coagulation have occurred following infusion of IGIV.

**Hemolysis (cont.):**

The following risk factors may be associated with the development of hemolysis following IGIV administration: High doses (e.g., 2 g/kg or more), given either as a single administration or divided over several days, and non-O blood group. Other individual patient factors, such as an underlying inflammatory state (as may be reflected by, for example, elevated C-reactive protein or erythrocyte sedimentation rate), have been hypothesized to increase the risk of hemolysis following administration of IGIV[1], but their role is uncertain.

Closely monitor patients for clinical signs and symptoms of hemolysis, particularly patients with risk factors noted above. Consider appropriate laboratory testing in higher risk patients, including measurement of hemoglobin or hematocrit.

If clinical signs and symptoms of hemolysis or a significant drop in hemoglobin or hematocrit have been observed, perform confirmatory laboratory testing, including direct antiglobulin test. If transfusion is indicated for patients who develop hemolysis with clinically compromising anemia after receiving ALYGLO (immune globulin intravenous, human-stwk), perform adequate cross-matching to avoid exacerbating ongoing hemolysis.

**Transfusion-Related Acute Lung Injury (TRALI):** Noncardiogenic pulmonary edema [Transfusion-Related Acute Lung Injury (TRALI)] may occur in patients administered ALYGLO[1]. TRALI is characterized by severe respiratory distress, pulmonary edema, hypoxemia, normal left ventricular function, and fever. Signs and symptoms typically appear within 1 to 6 hours following treatment. Patients with TRALI may be managed using oxygen therapy with adequate ventilator support.

Monitor patients for pulmonary adverse reactions. If TRALI is suspected, perform appropriate tests for the presence of antineutrophil antibodies and anti-human leukocyte antigen (HLA) antibodies in both the product and the patient's serum.

**Transmissible Infectious Agents:** Because ALYGLO is made from human blood, it may carry a risk of transmitting infectious agents, e.g., viruses, the variant Creutzfeldt-Jakob disease (vCJD) agent and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent. The risk of infectious agent transmission has been reduced by screening plasma donors and by including virus inactivation/removal steps in the manufacturing process of ALYGLO.

Report all infections thought by a physician possibly transmitted by ALYGLO to GC Biopharma USA, Inc. at 1-833-426-6426. Discuss the risks and benefits of its use with the patient before prescribing or administering this product.

**Monitoring Laboratory Tests**

- Periodic monitoring of renal function and urine output is particularly important in patients at increased risk of developing acute renal failure. Assess renal function, including measurement of blood urea nitrogen (BUN) and serum creatinine before the initial infusion of ALYGLO and at appropriate intervals thereafter.
- Because of the potential for increased risk of thrombosis with ALYGLO, consider baseline assessment of blood viscosity in patients at risk for hyperviscosity, including those with cryoglobulins, fasting chylomicronemia/markedly high triacylglycerols (triglycerides), or monoclonal gammopathies.
- If signs and/or symptoms of hemolysis are present after an infusion of ALYGLO, perform appropriate laboratory testing for confirmation.
- If TRALI is suspected, perform appropriate tests for the presence of anti-neutrophil antibodies in both the product and patient's serum.

**Interference with Laboratory Tests:** After infusion of immunoglobulin, the transitory rise of the various passively transferred antibodies in the patient's blood may yield positive serological testing results, with the potential for misleading interpretation. Passive transmission of antibodies to erythrocyte antigens (e.g., A, B, and D) may cause a positive direct or indirect antiglobulin (Coombs) test.

## ADVERSE REACTIONS

The most common adverse reactions, observed in ≥ 5% of study subjects, were headache, nausea/vomiting, fatigue, nasal/sinus congestion, rash, arthralgia, diarrhea, muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, and dizziness.

**Clinical Trials Experience:** Because clinical trials are conducted under widely varying conditions, adverse reaction rates observed in the clinical trials of a drug cannot be directly compared to rates in the clinical trials of another drug and may not reflect the rates observed in practice.

In an open-label, single-arm, multicenter, non-randomized clinical trial, 33 subjects with primary humoral immunodeficiency received doses of ALYGLO ranging from 319 mg/kg to 817 mg/kg every 21 days or 28 days for up to 12 months.

The passive transfer of antibodies with IGIV administration may interfere with the response to live virus vaccines such as measles, mumps, rubella, and varicella. Immunizing physicians should be informed of recent IGIV therapy so that appropriate measures may be taken.

Twenty-eight subjects (85%) experienced a total of 145 temporally associated adverse reactions (adverse events that occurred during or within 72 hours after the end of an infusion) during the study. The temporally associated ARs were headache (13 subjects, 39%), nausea/vomiting (11 subjects, 33%), fatigue (6 subjects, 18%), nasal/sinus congestion (5 subjects, 15%) rash (4 subjects, (12%), arthralgia, diarrhea (3 subjects, 9% each), muscle pain/aches, infusion site pain/swelling, abdominal pain/discomfort, cough, dizziness (2 subjects, 6% each).

These are presented in Table 2. There were no deaths and no adverse reactions leading to withdrawal from the study.

**Table 2 Adverse Reactions* (ARs) (within 72 hours after the end of an ALYGLO infusion) in ≥ 5% of Subjects**

| Adverse Reactions (ARs) | No. of Subjects Reporting ARs (Percentage of Subjects) [N[1]=33] | No. of Infusions with ARs (Percentage of Infusions) [N[2]=427] |
|---|---|---|
| Headache | 13 (39) | 32 (7.5) |
| Nausea/vomiting | 11 (33) | 20 (4.7) |
| Fatigue | 6 (18) | 18 (4.2) |
| Nasal/sinus congestion | 5(15) | 5 (1.2) |
| Rash | 4 (12) | 4 (0.9) |
| Arthralgia | 3 (9) | 4 (0.9) |
| Diarrhea | 3 (9) | 3 (0.7) |
| Muscle pain/aches | 2 (6) | 7 (1.6) |
| Infusion site pain/swelling | 2 (6) | 6 (1.4) |
| Abdominal pain/discomfort | 2 (6) | 3 (0.7) |
| Cough | 2 (6) | 2 (0.5) |
| Dizziness | 2 (6) | 2 (0.5) |

*Adverse events that occurred during or within 72 hours after the end of an infusion
[1]Total number of subjects
[2]Total number of infusions

**Postmarketing Experience:** Because postmarketing reporting of adverse reactions is voluntary and from a population of uncertain size, it is not always possible to reliably estimate the frequency of these reactions or establish a causal relationship to product exposure. The following adverse reactions have been identified and reported during the post-approval use of marketed IGIV products:

**Blood and lymphatic system disorders:** leukopenia, hemolysis, pancytopenia; **Immune system disorders:** hypersensitivity (e.g., anaphylaxis), anaphylactic shock, anaphylactic reaction, anaphylactoid reaction, allergic reaction, angioedema, face edema; **Metabolic and nutritional disorders:** fluid overload, (pseudo) hyponatremia; **Psychiatric disorders:** agitation, confusion, anxiety, nervousness; **Nervous system disorders:** coma, loss of consciousness, seizures, (acute) encephalopathy, cerebrovascular accident, stroke, aseptic meningitis, migraine, speech disorder, paresthesia, hypoesthesia, photophobia, tremor; **Cardiac disorders:** myocardial infarction, cardiac arrest, angina pectoris, tachycardia, bradycardia, palpitations, cyanosis; **Vascular disorders:** hypotension, (deep vein) thrombosis, peripheral circulatory failure/collapse, hypertension, phlebitis, pallor; **Respiratory, thoracic and mediastinal disorders:** apnea, Acute Respiratory Distress Syndrome (ARDS), TRALI, respiratory failure, pulmonary embolism, pulmonary edema, bronchospasm, dyspnea, hypoxia, wheezing, cough; **Gastrointestinal disorders:** diarrhea, hepatic dysfunction, abdominal discomfort; **Skin and subcutaneous tissue disorders:** eczema, urticaria, rash (erythematous), dermatitis, pruritus, alopecia, Stevens-Johnson syndrome epidermolysis, skin exfoliation, erythema (multiform), dermatitis (e.g., bullous dermatitis); **Musculoskeletal and connective tissue disorders:** back pain, arthralgia, myalgia, musculoskeletal pain, muscle stiffness, pain in extremity, neck pain, muscle spasm; **Renal and urinary disorders:** acute renal failure, osmotic nephropathy, renal pain; **General disorders and administration site conditions:** injection-site reaction, chills, chest pain or discomfort, hot flush, flushing, flu-like illness, feeling cold or hot, edema, hyperhidrosis, malaise, asthenia, lethargy, burning sensation; **Investigations:** hepatic enzymes increased, oxygen saturation decreased, falsely elevated erythrocyte sedimentation rate, positive direct antiglobulin (Coombs) test.

## DRUG INTERACTIONS

Clinical studies have not evaluated mixture of ALYGLO with other drugs and intravenous solutions. It is recommended that ALYGLO is administered separately from other drugs or medications which the patient may be receiving. Do not mix the product.

Transitory rise of the various passively transferred antibodies in the patient's blood after infusion of immunoglobulin may yield positive serological testing results, with the potential for misleading interpretation.

## USE IN SPECIFIC POPULATIONS

**Geriatric use:** In patients over age 65 or in any patient at risk of developing renal insufficiency, do not exceed the recommended dose, and infuse ALYGLO at the minimum infusion rate practicable.

**Reference: 1.** ALYGLO Prescribing Information. GC Biopharma USA, Inc.; 2023.


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# Advanced Certification:
## Level Up Your Infusion with a Certified IG Clinician

IG-certified clinicians improve the quality and safety of IG therapy, which directly enhances patient satisfaction and outcomes.

**By Rachel Colletta, BSN, CRNI, IgCN**

**IN THE HIGHLY** specialized field of immune globulin (IG) therapy, your infusion experience depends on the skills and qualifications of the clinicians who administer and manage your therapy. As a patient receiving IG therapy, you deserve to receive the highest level of care before, during and after your infusions. While many clinicians are well-qualified to administer IG therapy, having a healthcare team with certified clinicians is a way to ensure you receive the highest level of care.

The Immunoglobulin National Society (IgNS) offers the only certification in this field for nurses and pharmacists: the IG Certified Pharmacist (IgCP) and IG Certified Nurse (IgCN) designations. These certifications are more than just labels; they show excellence and advancement in this field and confirm the clinicians managing IG therapy have the knowledge and abilities to achieve the best patient outcomes.

IgNS certification highlights a healthcare professional's commitment to ongoing learning and excellence and enhances IG therapy safety and effectiveness. But how does certification work, who oversees the process and how does it benefit patients receiving IG therapy? This article will answer all of these questions and more.

## The Role of IgNS in Certifying Healthcare Professionals

IgNS plays a crucial role in standardizing the practice of IG therapy. At the heart of IgNS's objectives is the development of evidence-based standards of practice that ensure IG therapy is administered consistently and effectively, based on the latest scientific research and clinical evidence. By establishing these standards, IgNS helps clinicians deliver the highest level of care to patients receiving IG treatments. In addition to standards for clinicians, IgNS has developed and will soon release the *Guide to Immunoglobulin Therapy*, a resource for patients based on the same standards so they will have a better understanding of what to expect from certified clinicians when undergoing IG therapy.

Education is another critical component of IgNS's mission. IgNS offers various educational programs relevant to healthcare professionals such as nurses, pharmacists and physicians. These initiatives equip healthcare professionals with the knowledge and skills necessary to excel in IG therapy administration and management.

Another way IgNS champions best practices is through its professional certification programs for nurses and pharmacists. These certifications not only validate the specialized skills of clinicians in IG therapy but also serve as a mark of excellence in the field. By recognizing and promoting expertise, these certifications elevate the overall quality of care in IG therapy.

## The Certification Process

IgNS helps to ensure healthcare professionals are skilled in managing IG therapy. The certification process includes an exam that tests the knowledge and skills in various areas around IG therapy, including knowledge of disease states, IG products, administration, monitoring and follow-up, to name a few. Once the exam is passed, clinicians receive a three-year valid certification. During that time, they must earn recertification by attending conferences and keeping up to date with the latest clinical evidence and practices by taking additional educational courses.

This structured certification and recertification process not only validates the expertise of IG therapy clinicians but also helps to standardize IG practices within organizations and across sites of care. It empowers clinicians to make confident and competent clinical decisions, significantly improving patient outcomes. This certification serves as a measurable demonstration of expertise in IG therapy, which separates and elevates these clinicians from others.

## Advantages of Certified Nurses for Patients

Certified nurses with an IgCN credential contribute immensely to improving IG therapy management and enhancing patient care through their specialized knowledge and the ability to educate patients and families effectively. Some of the advantages of working with a certified nurse include:

- *Specialized knowledge in IG therapy.* Certification as an IgCN demonstrates advanced knowledge and competency in administering IG therapies. IG-certified nurses are trained in disease-specific indications, infusion techniques, patient monitoring and management of adverse reactions related to IG therapy. This enables them to deliver safe, effective, patient-centered care, enhancing treatment outcomes for individuals receiving IG therapy.
- *Validation of clinical competence.* IgCN certification validates the clinical competence of nurses in IG therapy management. It signifies that they have met advanced standards of practice and have the necessary skills to provide high-quality care. Certified nurses are skilled in assessing patient needs, implementing infusion protocols and responding promptly to any complications that may arise during therapy sessions.
- *Professional growth and recognition.* Obtaining IgCN certification enhances professional growth and recognition within the healthcare community. It demonstrates a commitment to continuous learning and adherence to best practices in IG therapy. Certified nurses are often recognized as leaders in their field, contributing to improved patient outcomes, enhanced team collaboration and a higher standard of care delivery across healthcare settings. Becoming certified means these clinicians have committed to continually advancing their knowledge and skills to improve the overall infusion experience for patients.

## Advantages of Certified Pharmacists for Patients

Having an IgCP on your healthcare team comes with substantial benefits. This certification signifies a pharmacist's advanced expertise and proficiency in managing IG therapies, encompassing important aspects such as disease-specific indications, dosing regimens, infusion techniques and adverse event management. Some of the advantages of working with a certified pharmacist include:

- *Enhanced expertise in IG therapy management.* Certification as an IgCP signifies specialized knowledge and skills in managing IG therapies. IgCPs are trained in disease-specific indications, dosing calculations, infusion protocols

and adverse event management related to IG therapy. This expertise allows them to develop personalized treatment plans to optimize therapy outcomes.

• *Recognition of professional credibility.* IgCP certification enhances the professional credibility of pharmacists in the field of IG therapy. It demonstrates their commitment to maintaining high standards of practice and staying updated with the latest advancements in IG therapies. This recognition boosts confidence among healthcare colleagues and instills patient trust, improving patient outcomes and satisfaction.

• *Contribution to interdisciplinary collaboration.* IgCPs play a vital role in interdisciplinary healthcare teams by bridging gaps between pharmacy and clinical practice. They work closely with physicians, nurses and other healthcare professionals to effectively coordinate patient care. IgCPs contribute to comprehensive patient management and ensure standardized care delivery across healthcare settings by sharing their specialized knowledge and participating in treatment decisions.

**Certified nurses with an IgCN credential contribute immensely to improving IG therapy management and enhancing patient care through their specialized knowledge and the ability to educate patients and families effectively.**

## Enhanced Quality of Care

Nurses and pharmacists who achieve the IgCN and IgCP credentials significantly enhance the quality of IG therapy practice for patients. By standardizing practices, these credentials ensure all procedures and treatments meet high standards, directly improving patient care, outcomes and satisfaction.

The credentialing of nurses and pharmacists fosters improved trust and confidence among patients and prescribers, which is crucial for effective communication and patient engagement. Certified professionals are seen as empathic and competent partners in the patient's health journey.

Communication skills training for these professionals further enhances their ability to understand and address patient concerns, making the healthcare experience more personalized and supportive. This not only improves patient satisfaction but also creates a deeper connection between the patient and the provider, enhancing adherence to treatment plans and overall care effectiveness.

## Call to Action

IgCNs and IgCPs play a crucial role in improving the quality and safety of IG therapy, which directly enhances your satisfaction and outcomes. Through a rigorous certification process, these healthcare professionals gain essential knowledge and skills that make them integral to managing IG therapy effectively. Certified clinicians focus on patient-centered care, ensuring the therapy you receive is well-tolerated and fits your lifestyle and preferences. They excel in creating personalized therapy plans based on your tolerance of therapy and current medical conditions. Certified clinicians are competent in educating you on your therapy, handling adverse reactions and ensuring top-notch care that focuses on your well-being.

As healthcare evolves, the expertise of IG-certified professionals strengthens relationships between you and your healthcare team, fostering trust and excellence in patient care. Better patient outcomes highlight the importance of supporting and encouraging your healthcare team to become certified in IG therapy.

Ask your nurse and pharmacist if they are IG-certified to ensure you receive the best care possible. This proactive step can significantly enhance your infusion experience and improve the overall success of your treatment plan.

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**RACHEL COLLETTA**, BSN, CRNI, IgCN, is director of educational resources at the Immunoglobulin National Society.

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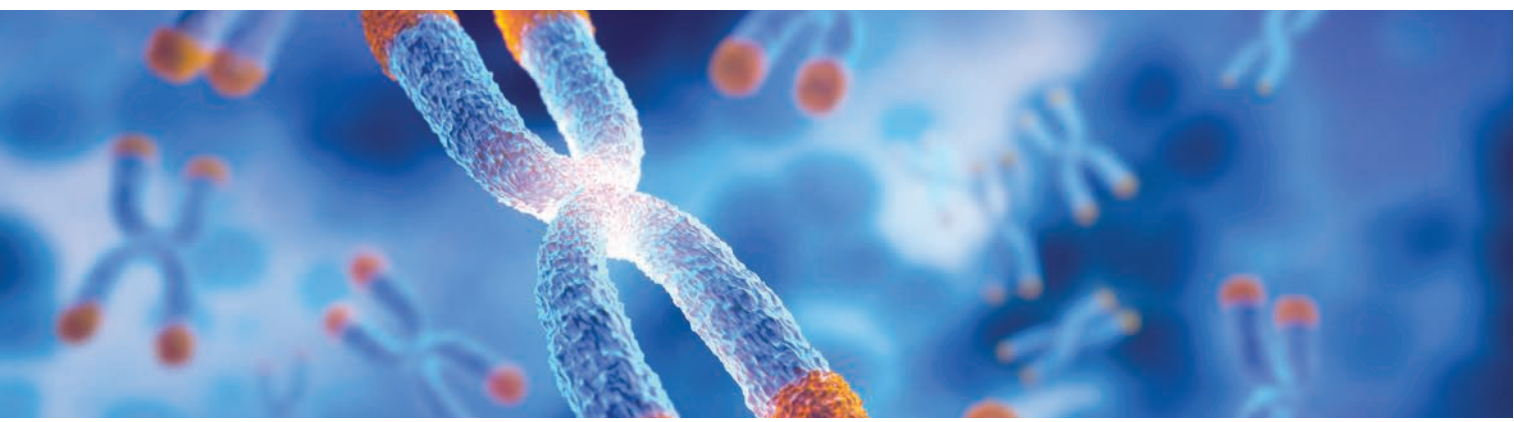
# X-Linked Carriers: What Everyone Needs to Know

While most people are aware of the concept of recessive genes, which can result in medical problems , they are not as aware of X-linked recessive conditions, for which females can be a carrier and the disease can be expressed in male offspring. Fortuntely, most remain rare, but many can be now tested for.

**By Jim Trageser**



**MOST PEOPLE HAVE** a basic understanding of recessive vs. dominant genes based on high school biology lessons involving eye color: Brown eyes are the result of a dominant gene and blue eyes are the result of a recessive gene. So a child with blues eyes had to get the blue eye gene from both parents.

While recent research has shown that eye color is based upon multiple genetic factors — and that, in fact, two blue-eyed parents can have a brown-eyed baby[1] (a situation that in previous generations led to accusations of adultery) — the basic premise remains: The human genome is comprised of 46 total chromosomes, matched in 22 pairs, with females having two X chromosomes and males having an X chromosome and a Y chromosome, and the 23rd set, with both mom and dad each contributing half, under normal circumstances, to each child. Every cell in our bodies has chromosomes in its nucleus (except red blood cells and platelets, which lack nuclei). Each chromosome is made up of genes — DNA molecules containing coded instructions on everything from how to build new cells to how to regulate biochemical reactions.

The 23 pairs of chromosomes range in size, from the largest first pair, in which each chromosome has nearly 3,000 genes, to smaller pairs averaging about 1,000 genes each. However, the truncated Y chromosome on the 23rd pair is much smaller than its X chromosome counterpart, with the X chromosome comprised of about 900 genes, and the Y between 70 and 200.[2]

Since males always get the Y chromosome from their father, and since the Y chromosome is missing many genes contained on the X chromosome from their mother, even a normally recessive gene on the X chromosome can become dominant if it is not part of the Y chromosome. All of this means that X-linked recessive disorders are far more likely to affect males. Further, females can carry the gene without knowing it, and are known as obligate carriers.

## Dominant vs. Recessive

In every chromosome pair, there are normally two versions of each gene. These are known as alleles. Thus, we have a chromosome from each parent to generate the pair and, therefore, typically at least two alleles of each gene, again, one from each parent. A dominant gene is one that has precedence over the other.[3] So, when a dominant gene is expressed, it is the active gene. If it contains a deleterious mutation, this can create a medical problem.[4]

In contrast, recessive gene mutations must be present

on both alleles to express a medical condition. This means each parent must be carrying "one" gene with a deleterious mutation, and they must pass that gene from each parent into an offspring. The parents are not affected since they each have one functional gene copy, but the offspring will be affected, since they receive only two copies of genes that cannot carry out normal function.

An easy-to-understand example of a recessive gene is the melanocortin-1 receptor (MC1R) gene, found on the 16th pair of chromosomes. This gene regulates the production of melanin — the pigment that determines our skin tone and hair color (and how well we tan). There is an altered version of this gene in which the body does not dispose of red pigment. This version of the gene is recessive, meaning most people who have the gene will not have red hair because the other gene still instructs the body to get rid of red pigment. If both parents contribute the altered gene, however, the child will have red hair.[5]

### What It Means to Be X-Linked Recessive

The X chromosome is unique since recessive genes can only be passed on from the mother. Any male with a genetic abnormality on the X gene will have the associated condition related to the faulty gene since there is no matching gene on the Y chromosome to counter it. By definition, then, only females can carry a recessive gene on the X chromosome.

This also means that a mother who does not exhibit any symptoms or signs of a genetic disorder can still pass them on to her son, and her daughters will have a 50 percent chance of carrying that recessive gene themselves.[6] (However, if a son does not develop the condition, then he did not inherit the gene and cannot pass it on to future generations.[7]) Each time she conceives, a mother has a 50 percent chance each of a son with the gene, a son without the gene, a daughter carrying the gene and a daughter without the gene (Figure).

The X chromosome has more than 500 genetic disorders associated with its genes already. The genes on the X chromosome are often tied to the building of various tissues: blood, bone, nerves, heart, skin, etc. Thus, genetic abnormalities on the X chromosome tend to lead to significant health challenges in those who inherit them.[8]

### X-Linked Recessive Conditions

Numerous conditions are tied to genetic mutations on the X chromosome. Some of the more well-documented and studied include:

*Adrenoleukodystrophy (ALD).* Caused by an abnormality on the ABCD1 gene, ALD leads to a breakdown in the myelin sheath that covers the brain, spinal cord and nerve cells. Symptoms include weakening of muscles, spasms, seizures, hearing loss and/or difficulty swallowing.[9] While males are most severely affected, even female carriers can develop mild symptoms in their adult years (a condition known as adrenomyeloneuropathy).

*Aldred syndrome.* Tied to a missing gene on the p11.3 area of the X chromosome, Aldred syndrome leads to moderate learning disability and visual impairment.[10]

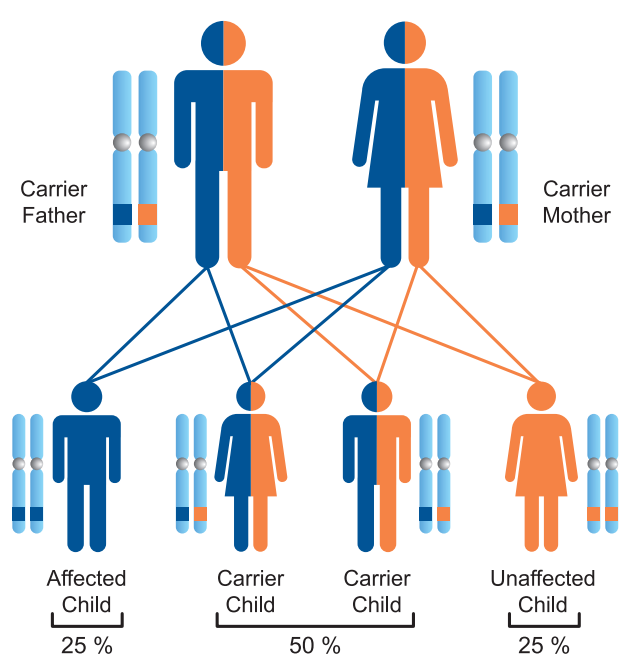
*Alport syndrome.* Mutations in the COL4A3-A5 genes lead to an inability of the body to correctly make collagen (only the COL4A5 gene is on the X chromosome; the other two variants are non-X-linked). The primary symptom is kidney disease, as the collagen is used to filter the blood, but hearing and vision issues can also arise.[11]

*Barth syndrome.* A defective TAZ gene on the X chromosome leads to an enlarged heart, weakened muscles and frequent infections.[12]

*Becker muscular dystrophy (BMD).* Caused by a mutation on the dystrophin gene, BMD leads to a progressive weakening of the leg muscles.[13]

*Charcot-Marie–Tooth disease (CMT).* Normally caused by a mutation on chromosome 17, some versions are X-linked.

**Figure. Autosomal Recessive Inheritance**



HyQvia

[Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase]


## What is HyQvia®?

HyQvia [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase] is a liquid medicine that is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years and older.

## IMPORTANT SAFETY INFORMATION

### What is the most important information that I should know about HyQvia?

- HyQvia can cause blood clots.
- Call your healthcare professional (HCP) if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
- Your HCP may perform blood tests regularly to check your IgG level.
- Do not infuse HyQvia into or around an infected or red swollen area because it can cause infection to spread.

### Who should not take HyQvia?

Do not take HyQvia if you:

- Are allergic to IgG, hyaluronidase, other blood products, or any ingredient in HyQvia.

### What should I avoid while taking HyQvia?

- HyQvia can make vaccines (like measles/mumps/rubella or chickenpox vaccines) not work as well for you. Before you get any vaccines, tell your HCP that you take HyQvia.

### What should I tell my HCP before I start using or while using HyQvia?

Tell your HCP if you:

- Have or had any kidney, liver, or heart problems or history of blood clots because HyQvia can make these problems worse.
- Have IgA deficiency or a history of severe allergic reactions to IgG or other blood products.
- Are pregnant, trying to become pregnant or are breast feeding. It is not known whether HyQvia can harm the unborn baby or breastfed infant.

### What are the possible or reasonably likely side effects of HyQvia?

**HyQvia can cause serious side effects. If any of the following problems occur after starting HyQvia, stop the infusion immediately and contact your HCP or call emergency services:**

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.

# Meet the only monthly* subQ IG treatment and say hy to more of what you love.

## **0.025 infections** per year

This is equivalent to 25 acute serious bacterial infections (ASBIs) out of 1,000 patients over the course of the 12-month study period.

The FDA standard for efficacy—that is, if an immunoglobulin works—is less than 1 ASBI per year. In the clinical trial, people taking HyQvia experienced significantly less than that.

## **0 days** in the hospital per year

There was a mean of 0.037 days spent in the hospital due to infection during the study.

## **<4 days** off work or school per year

On average, patients taking HyQvia missed 3.31 days of work or school due to an infection.

- HyQvia was studied in a clinical trial of 83 people with PI, with the main goal of measuring how many acute serious bacterial infections (ASBIs) they experienced over the course of 1 year
- ASBIs are short-term but serious infections caused by bacteria that require immediate medical care
- ASBIs included 2 episodes of pneumonia, both treated as outpatients with oral antibiotics. An additional episode of pneumonia requiring hospitalization occurred during the ramp-up
- The most common general (systemic) side effects were headache, antibody formation against hyaluronidase (Hy), fatigue, nausea, fever, and vomiting. The most common side effects at the infusion site (local) were pain, redness, swelling, and itching

*Between infusions, based on administration every 3 or 4 weeks.
subQ IG=subcutaneous immune globulin.

## IMPORTANT SAFETY INFORMATION (continued)

- Chest pain or trouble breathing, blue lips or extremities. These could be signs of a serious heart or lung problem.
- Fever over 100°F. This could be a sign of an infection.

After HyQvia infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused. The following possible side effects may occur at the site of infusion and generally go away within a few hours, and are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Swelling
- Itching

The most common side effects of HyQvia are:

- Headache
- Fatigue
- Nausea
- Fever
- Vomiting

Antibodies to the hyaluronidase component of HyQvia were formed in some patients taking HyQvia. It is not known if there is any long-term effect. In theory, these antibodies could react with your body's own hyaluronidase (PH20). PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side-effects.

**These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.**

***Please see Important Facts about HyQvia on the following page.***

**You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-FDA-1088.**

**Up to 100% of out-of-pocket co-pay costs could be covered.**

Scan the QR code to learn more about HyQvia, including co-pay costs.



Takeda

**IMPORTANT FACTS about HYQVIA (Hi-Q-via) [Immune Globulin Infusion 10% (Human) with Recombinant Human Hyaluronidase] Solution, for subcutaneous administration**

## What is the most important information I should know about HYQVIA?

- HYQVIA can cause blood clots.
- Call your healthcare provider (HCP) if you have pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s), unexplained shortness of breath, chest pain or discomfort that worsens on deep breathing, unexplained rapid pulse, numbness or weakness on one side of the body.
- Your HCP may perform blood tests regularly to check your IgG level.
- Do not infuse HYQVIA into or around an infected or red swollen area because it can cause infection to spread.

## What is HYQVIA?

HYQVIA is a liquid medicine containing immune globulin and Recombinant Human Hyaluronidase. HYQVIA is given under the skin (subcutaneously) to treat primary immunodeficiency (PI) in people 2 years of age and older. HYQVIA contains IgG antibodies, collected from human plasma donated by healthy people.

- The antibodies help your body to fight off bacterial and viral infections.
- The hyaluronidase is found in your body naturally. It's the first part of your two-part infusion. It temporarily opens the space under your skin (the subcutaneous space), allowing a larger amount of IgG to reach your subcutaneous tissue and be absorbed into your bloodstream.

## What should I tell my HCP before I start using or while using HYQVIA?

Tell your HCP if you:

- Have or had any kidney, liver, or heart problems or history of blood clots because HYQVIA can make these problems worse.
- Have IgA deficiency or a history of severe allergic reactions to IgG or other blood products.
- Are pregnant, trying to become pregnant, or are breastfeeding. It is not known whether HYQVIA can harm the unborn baby or breastfed infant.

## Who should not take HYQVIA?

- Do not take HYQVIA if you are allergic to IgG, hyaluronidase, other blood products, or any ingredient in HYQVIA.

## How should I take HYQVIA?

- HYQVIA is infused under the skin (subcutaneously) up to once every 4 weeks.
- You can get HYQVIA at your HCP's office, clinic, or hospital.
- You can use HYQVIA at home. You and your HCP will decide if home self-infusion is right for you.

## What are the possible or reasonably likely side effects of HYQVIA?

After HYQVIA infusion a temporary, soft swelling may occur around the infusion site, which may last 1 to 3 days, due to the volume of fluid infused.

The following local reactions may occur at the site of infusion and generally go away in a few hours. Local reactions are less likely after the first few infusions.

- Mild or moderate pain
- Redness
- Swelling
- Itching

The most common side effects of HYQVIA are: headache, fatigue, nausea, fever, and vomiting.

Antibodies to the hyaluronidase component of HYQVIA were formed in some patients taking HYQVIA. It is not known if there is any long-term effect. In theory, these antibodies could react with your body's own PH20. PH20 is present in the male reproductive tract. So far, these antibodies have not been associated with increased or new side effects.

Call your HCP or go to your emergency department right away if you get:

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting or dizziness. These could be signs of a serious allergic reaction.
- Bad headache with nausea, vomiting, stiff neck, fever, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain.
- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem.
- Pain, swelling, warmth, redness, or a lump in your legs or arms, other than at the infusion site(s). These could be signs of a blood clot.
- Brown or red urine, fast heart rate, yellow skin or eyes. These could be signs of a liver or blood problem.
- Chest pain or trouble breathing, blue lips or extremities. These could be signs of a serious heart or lung problem.

These are not all of the possible side effects for HYQVIA. You can ask your HCP for information that is provided to HCPs. Talk to your HCP about any side effects that bother you or that don't go away.

## How do I store HYQVIA?

Store HYQVIA refrigerated or at room temperature.

- You can store HYQVIA in the refrigerator (36°F to 46°F [2°C to 8°C]) for up to 36 months.
- You can store HYQVIA at room temperature (up to 77°F [25°C]) for up to 3 months during the first 24 months from the date of manufacturing (Mfg Date) printed on the carton.
- Do not return HYQVIA to the refrigerator if you take it out to room temperature.

Check the expiration date on the carton and vial label. Do not use HYQVIA after the expiration date.

Do not freeze.

Protect from light. You can use the original HYQVIA containers to protect it from light.

## How do I get more information about HYQVIA?

The risk information provided here is not comprehensive. To learn more, talk about HYQVIA with your HCP or pharmacist. The FDA-approved Full Prescribing Information, including Information for Patients, can be found at www.HYQVIA.com or by calling 1-877-TAKEDA7 (1-877-825-3327).



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US-HYQ-0299v5.0 02/24

The disease is marked by muscle weakness in the arms and legs, and decreased nerve sensitivity.[14]

*Coffin-Lowry syndrome (CLS).* This is a very rare condition sometime caused by a defective RPS6KA3 gene. Other cases are sporadic, meaning the cause is unknown. Symptoms include skeletal and facial abnormalities, developmental slowness and cardiovascular issues.[15]

*DDX3X syndrome.* One of the more recently X-linked genetic diseases, this condition has so far been observed more often in females than males, although researchers do not yet understand why. It is linked to autism spectrum disorder, attention-deficit/hyperactivity disorder and language-learning delays.[16]

*Duchenne muscular dystrophy (DMD).* As with BMD, DMD is caused by a defective dystrophin gene. It is the most severe of the inherited muscular dystrophies.[17]

*Endocardial fibroelastosis.* Recent studies suggest an X-recessive cause to this condition that leads to an enlarged heart.[18]

*Fabry disease (also known as Anderson-Fabry disease).* The result of a defective GLA gene, Fabry disease results in an accumulation of fats in cells, resulting in cardiovascular and renal disease.[19]

*FG syndrome.* An abnormal MED12 gene can lead to learning disability, facial deformities, incomplete brain development and anal abnormalities.[20]

*Fragile X syndrome (also known as Martin-Bell syndrome).* A mutation in the FMR1 gene prevents the proper development of synapses. This is the most common cause of autism spectrum disorder.[21]

*Hemophilia A.* Caused by a defective HEMA gene, patients with this disease have blood that is unable to form clots, leading to even minor cuts and bruising causing excessive bleeding.[22]

*Hypohidrotic ectodermal dysplasia (HED).* Most cases of HED are tied to a mutated EDA gene on the X chromosome. The disease is marked by a radically reduced ability to sweat, missing teeth and thin scalp and hair.[23]

*Lesch-Nyhan syndrome.* Caused by a mutated HPRT1 gene that regulates metabolism of broken down DNA and RNA compounds, Lesch-Nyhan syndrome is marked by arthritis, poor muscle control and a strong tendency toward self injury.[24]

*MASA syndrome.* This syndrome, linked to a mutated L1CAM gene, causes developmental disability, delayed speech, difficulty walking and enlarged ventricles in the heart.[25]

*Nasodigitoacoustic syndrome (Keipert syndrome).* Tied to a mutated GPC4 gene, this condition is marked by skeletal, facial and heart deformities, as well as learning disabilities.[26]

*Norrie disease.* A result of a defective NDP gene, Norrie disease prevents the retina from forming properly, leading to blindness.[27]

*Occipital horn syndrome (OHS).* Caused by a mutated ATP7A gene, OHS is an extremely rare condition that prevents the body from properly processing copper. Symptoms include calcium deposits on the skull, loose skin and joints, mild learning disability, and degraded nerves controlling nonvoluntary body functions.[28]

*Red-green color blindness.* One of the most common and least-serious X-linked conditions, this is caused by a mutation on the OPN1LW or OPN1MW gene.

## Starting a Family

The above list of X-linked conditions can be intimidating, if not frightening. However, when considering to have children, the reality is that most of these diseases remain rare. Unfortunatly, the family history may not be helpful in knowing an X-linked recessive condition exists, and one-third of all X-linked recessive conditions are due to a "new" mutation (the Haldane hypothesis from 1935), resulting in X-linked recessive diseases being maintained at constant levels in the general population. Additionally, for X-linked severe combined deficiency, the "de novo" mutation most likely occurred in the maternal grandfather of the affected male child.

It is important to know your family medical history. While discussing genetic diseases can be difficult, usually there is someone — a cousin, a great-uncle — who is willing to talk about it if immediate family members are uncomfortable or unwilling.

A genetic counselor can be a wonderful asset in this process. The counselor can help review your family history, assist in planning genetic testing and walk you through decision-making.[29] Your insurance may even cover all or part of the cost of working with a genetic counselor. The National Society of Genetic Counselors can help you navigate that landscape as well. The Society hosts two websites to help families: findageneticcounselor.nsgc.org and www.aboutgeneticcounselors.com.

Due to the wide variety of diseases tied to recessive gene mutations on the X chromosome, there is no single genetic test to look for all of them. Most would have to be tested for separately, and not all insurance plans will cover these.

But again, in most families, if there is a suspicion, it is usually about a specific disease, and a genetic test can be performed for any of those diseases listed above.

## What It All Means

Genes can be either dominant or recessive, and because of random chance mutations, genes differ and not all promote survival. Genetic mutations on the X chromosome can lead to numerous conditions, which are typically recognized by most family trees. If there is a known X-linked recessive disorder in the family history, every family will have to come to its own decision about whether to attempt a pregnancy. Genetic testing can offer greater clarity, and genetic counseling can allow a couple to make as fully informed a decision as possible.

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**JIM TRAGESER** is a freelance journalist in the San Diego, Calif. area.

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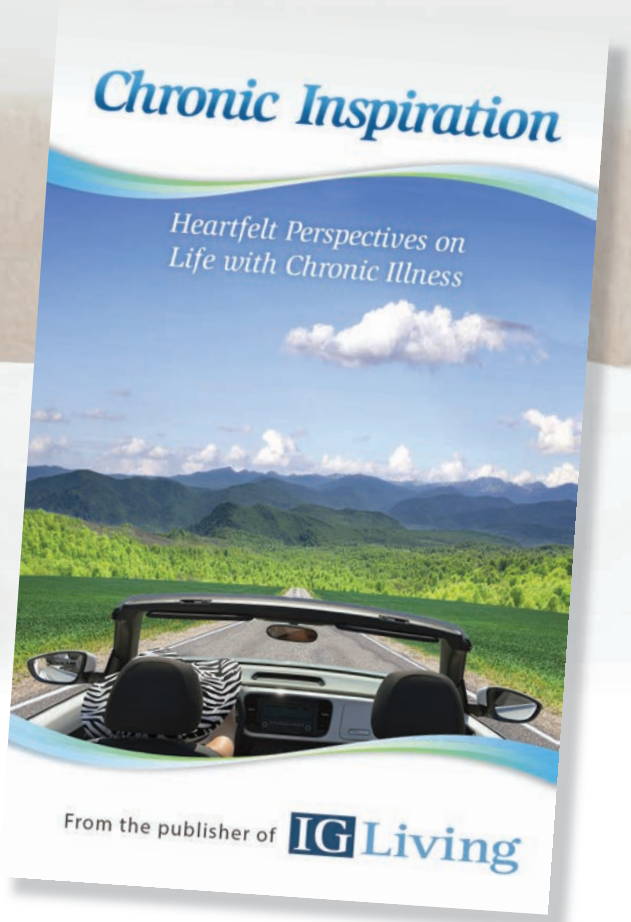


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*— Jenny Gardner*

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# *Profile:* Megan Anne Ryan

By Trudie Mitschang

*Since Megan Ryan's diagnosis of CVID, she led a demanding career and now makes it her life to work with patients and advocate for the PI community.*

**MEGAN ANNE RYAN** serves as a voice for patients worldwide. She is a proud native Texan who was diagnosed with common variable immune deficiency (CVID) in her 20s. Since then, Megan has become an outspoken and highly visible patient advocate, speaking at national conferences. Her unique perspective serves to shed light on the importance of plasma donors and the staff involved in plasma collection. Megan also served as the board treasurer for the Jose Antonio Grifols Lucas I Foundation and has further expanded her reach to the organization Undies for Everyone. She is a frequent contributing writer for *IG Living.*

**Trudie:** Can you tell us the story about your diagnosis?

**Megan:** Let me take you back more than 20 years ago. I was a recent college graduate beginning my career, and I had just married my husband. In the summer of 2001, I was diagnosed with CVID. I was thankful for a rather quick diagnosis and that a treatment option was available. I was originally seen by a primary care physician as a part of a routine physical to determine why I was having muscle pain in my lower legs. Laboratory results came back inconclusive, so she did more and more testing. At that time, I did not know her practice was focused on serving people living with HIV and AIDS, so she knew quite a bit about the immune system. Upon receipt of more conclusive tests, she indicated I was out of her scope of practice, and I am thankful that she referred me to a clinical immunologist who quickly took my case and confirmed my diagnosis with further testing. I was lucky because many patients can go years without a formal diagnosis.

**Trudie:** How did you navigate this life-changing news?

**Megan:** Upon diagnosis, my immunologist specifically advised me to focus on the positive. He emphasized that a treatment option was available and assured me that the treatment was going to be successful. I heeded his advice and focused on the positive from the beginning. Of course, there have been some low points and unexpected stops on my journey, but I've learned the power of resilience and that I can bounce back from that low point. Over the past two decades, even as I strive to live a very healthy lifestyle through diet, exercise, good mental health practices and being very compliant with my treatment plan, I've learned to say no at times to manage my energy levels and accept help when offered by others. I've learned I can bounce back.

**Trudie:** Tell us about your infusion plan.

**Megan:** When I first began immune globulin (IG) treatment, I would have to take time off work and go to a hospital infusion center to spend much of the day with an IV in my arm. Today, I receive my infusions through a portable infusion pump about the size of my hand. It lets me infuse IG on the go. I infuse every week without fail, and the entire process takes about two-and-a-half hours. My ability to do infusions wherever I am enables me to travel the country and help educate patients and healthcare providers about primary immunodeficiency (PI) diseases. Thanks to the advancement of medicine, my healthcare team, the legion of plasma donors and the support of my community and my advocacy work, two decades later, I'm full of hope for the future.

**Trudie:** How did you initially get involved in advocacy work?

**Megan:** I learned about the Immune Deficiency Foundation in 2008. Since then, I've worked with the organization as a patient advocate to help educate patients and healthcare providers about PI and the importance of plasma donation to make therapies for patients.

I currently serve as a peer-support coach and support group leader working to meet patients where they are on their journey. I also speak at plasma donation centers where I thank donors and put a face to who is helped by their plasma donations, making that human-to-

human connection. I also like to help employees at plasma donation centers find deeper meaning in the work they do by sharing my story.

Additionally, I've been involved in federal advocacy efforts and was the sole U.S. contributor to the International Patient Organisation for Primary Immunodeficiencies Learning Expedition to identify, analyze and provide recommendations on issues brought about by the COVID-19 pandemic for patients with PI. This report was published in January 2021 to build awareness of the physical, social and emotional well-being needs of patients in our community.

**Trudie:** You faced some recent financial challenges. Tell us about that.

**Megan:** Over the years, I had paid close to nothing out of pocket for my IG medication thanks to co-pay assistance programs. Then, I was unexpectedly hit with over $6,000 in medical bills. I spent two days working to uncover why I was receiving unexpected medical bills, and I learned I had a "specialty co-pay offset benefit" on my pharmacy plan that ensures I get *no* "benefit" from co-pay assistance programs! In my hours on the phone, I worked to understand why none of the IG product charges were being applied to my deductible or co-insurance. Financial summaries on the pharmacy website read like a foreign language and showed that charges for the product were almost zeroed out, as though they did not even exist, but I could see payments made by the co-pay assistance programs.

Here's what I learned: Pharmacy benefit managers work to negotiate these savings that they pocket directly — to the harm of patients. So, while the insurance company is paid by the third party, the patient is charged as well.

This is an insurance practice known as "co-pay accumulator programs," and patients find themselves slapped with a surprise bill when they uncover none of these payments have been applied to their deductibles or out-of-pocket costs. That's what happened to me. This experience expanded my advocacy work to help ban co-pay accumulators so others can avoid this experience.

**Trudie:** How have you lived a life of resilience?

**Megan:** I spent almost 22 years working in a demanding professional services environment where I contributed to the strategy and operational leadership of a large business unit. During many Friday afternoon meetings, I would infuse my IG product with few people knowing. In the workplace, I was never hesitant to share my diagnosis or journey as a person living with a rare and chronic disease. Today, I apply that professional experience serving on three nonprofit boards and a few other advisory committees. I now spend more time focused on advocacy in our patient community working with newly diagnosed patients, caregivers and healthcare providers. I've taken my weekly treatments on the road to many of the 50 states, more than 20 countries and four continents so far.

**Trudie:** What do you wish people understood about chronic illness?

**Megan:** Not all people living with physical limitations outwardly show their illness or limitations. The world is filled with people living with chronic pain or others living with potentially debilitating fatigue. And while they live with the illness associated with these symptoms, they also live with potential anxiety, guilt, depression, exhaustion, frustration and sometimes isolation. Like many others, I am challenged each time a person comments to me: "But you don't look sick." That phrase can invalidate a person living with an invisible illness. Some days, it takes significant effort to generate an outward appearance to project to the world that I have it all together.

**Trudie:** What advice would you give to other people who have disabilities or limitations?

**Megan:** Your medical diagnosis(es) should not define who you are as a person. I am not Megan with CVID — I am far more than that. I am a community volunteer, nonprofit leader, experienced financial operations professional, talented communicator, patient advocate, passionate friend, wife and daughter. I love to dig in the dirt and grow beautiful plants and flowers, and I'm always planning my next travel adventure!

TRUDIE MITSCHANG is a contributing writer for *IG Living* magazine.


2024 National Immunoglobulin (Ig) Treatment Survey
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Do you have a primary immunodeficiency?
Do you utilize Ig therapy?
Share your experiences with Ig treatment in our survey to help shape better care and support.
Your insights can make a difference!
Scan the QR code to see if you qualify
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A COMMUNITY EMPOWERED


Whether you've been recently diagnosed, have been living with a primary immunodeficiency (PI) for years, or just think you might have a PI, The Immune Deficiency is here to help.

While PI has no cure, there are lifesaving treatments available that can improve your quality of life. Our programs are meant to connect, engage, and empower families to live longer, stronger, healthier lives.

SCAN ME

# Immune Deficiency Foundation

primaryimmune.org | idf@primaryimmune.org


## INDICATIONS AND USAGE

PANZYGA (Immune Globulin Intravenous [Human] – ifas) is indicated for the treatment of primary humoral immunodeficiency (PI) in patients 2 years of age and older, chronic immune thrombocytopenia (cITP) in adults and chronic inflammatory demyelinating polyneuropathy (CIDP) in adults.

PANZYGA is a liquid medicine for infusion that contains immunoglobulin G (IgG), which are proteins that help fight infection. It is made from human plasma that is donated by healthy people and contains antibodies. For patients with PI, PANZYGA helps replace the missing antibodies in the body. For patients with cITP, PANZYGA helps the body produce more platelets (the blood cells that help blood clot) to control or prevent bleeding. For patients with CIDP, PANZYGA may help improve mobility and hand strength.

PANZYGA is given into a vein (intravenously) in a hospital, infusion center, doctor's office, or at home by a trained healthcare provider (HCP).

## IMPORTANT SAFETY INFORMATION

**WARNING: THROMBOSIS, RENAL DYSFUNCTION, and ACUTE RENAL FAILURE**

**See full prescribing information for complete BOXED WARNING**

- **Thrombosis may occur with immune globulin intravenous (IGIV) products, including PANZYGA. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **Renal dysfunction, acute renal failure, osmotic nephropathy, and death may occur with the administration of IGIV products in predisposed patients. Renal dysfunction and acute renal failure occur more commonly in patients receiving IGIV products containing sucrose. PANZYGA does not contain sucrose.**
- **For patients at risk of thrombosis, renal dysfunction, or acute renal failure, administer PANZYGA at the minimum infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**

**Do not use PANZYGA if you:**

- Have had a severe allergic reaction to immune globulin or other blood products
- Have a condition called selective (or severe) immunoglobulin A (IgA) deficiency, with antibodies against IgA and a history of hypersensitivity

**What should I know before taking PANZYGA?**

- PANZYGA can make vaccines (like measles/mumps/rubella or chickenpox vaccines) work less effectively for you. Before you get any vaccines, tell your healthcare provider that you take PANZYGA
- Decreased kidney function and kidney function failure can occur
- Severe headache, drowsiness, fever, painful eye movements, or nausea and vomiting can occur
- Elevated blood pressure can occur particularly in patients who have a history of hypertension (high blood pressure)
- If you are elderly, with heart or kidney problems, discuss with your healthcare provider prior to initiating treatment with PANZYGA
- PANZYGA is made from human blood and therefore may have a risk of transmitting infectious agents, including viruses and, theoretically, the variant Creutzfeldt-Jakob disease (CJD) and CJD agent. The production and manufacturing process reduces this risk, but the risk cannot be eliminated

**PANZYGA can cause serious side effects. If any of the following problems occur after starting PANZYGA, stop the infusion immediately and contact your HCP or call emergency services:**

- Hives, swelling in the mouth or throat, itching, trouble breathing, wheezing, fainting, or dizziness. These could be signs of a serious allergic reaction
- Bad headache with nausea, vomiting, stiff neck, fever, drowsiness, painful eye movements, and sensitivity to light. These could be signs of irritation and swelling of the lining around your brain

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**FDA approved for chronic inflammatory demyelinating polyneuropathy (CIDP) in adults to improve neuromuscular disability and impairment**

- **80% treated with 1g/kg and 92% treated with 2g/kg of PANZYGA saw improvement in arm and/or leg impairment***
- **With the PANZYGA Co-Pay Program, eligible patients may pay as little as $0 for PANZYGA†**
  - Patients must have commercial insurance to be eligible
  - Patients are not eligible if they are enrolled in a state or federally funded insurance program

*Depending on the ongoing therapy dose.
†Eligible, commercially insured patients may pay as little as $0 for PANZYGA and may receive a maximum benefit of $12,500 per year or the cost of patient's co-pay in a 12-month period (whichever is less) for claims received by the program. Terms and conditions/eligibility requirements apply. See full Terms and Conditions at PanzygaCoPay.com.

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**Talk to your doctor about PANZYGA and learn more at PanzygaInfo.com**

**IMPORTANT SAFETY INFORMATION (continued)**

- Reduced urination, sudden weight gain, or swelling in your legs. These could be signs of a kidney problem (decreased kidney function or kidney failure)
- Pain, swelling, warmth, redness, or a lump in your legs or arms. These could be signs of a blood clot, which could happen in the heart, brain, lungs, or elsewhere in the body
- Brown or red urine, swelling, fatigue, fast heart rate, difficulty breathing, or yellow skin or eyes. These could be signs of a liver or blood problem
- Chest pain or trouble breathing, or blue lips or extremities. These could be signs of a serious heart or lung problem
- Fever over 100°F. This could be a sign of an infection
- Headache, fatigue or confusion, vision problem, chest pain, difficulty breathing, irregular heartbeat, or pounding in your chest, neck, or ears. These could be signs of high blood pressure

Ask your HCP whether you should have rescue medications available, such as antihistamines or epinephrine.

**What are the possible or reasonably likely side effects for PANZYGA?**

The most common side effects that may occur with PANZYGA are:

- Headache
- Nausea
- Fever
- Increased blood pressure
- Dermatitis
- Fatigue
- Abdominal pain
- Dizziness
- Anemia

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away.

Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing.

**Patients should always ask their doctors for medical advice about adverse events.**

**You may report an adverse event related to Pfizer products by calling 1-800-438-1985 (US only). If you prefer, you may contact the U.S. Food & Drug Administration (FDA) directly. The FDA has established a reporting service known as MedWatch where healthcare professionals and consumers can report problems they suspect may be associated with the drugs and medical devices they prescribe, dispense, or use. Visit www.fda.gov/MedWatch or call 1-800-FDA-1088.**



## PANZYGA is FDA approved for 3 indications:

CIDP in adults
PI in patients 2 years of age or older
cITP in adults

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PP-PAN-USA-0927

April 2024

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Immune Globulin
Intravenous (Human) - ifas
10% Liquid Preparation

## CONSUMER BRIEF SUMMARY

(PANZYGA: *pan-zee-guh*)

This brief summary highlights the most important information about PANZYGA. Please read it carefully before using PANZYGA and each time you have an infusion, as there may be new information. This brief summary does not take the place of talking with your healthcare provider about your medical condition or your treatment. If you have any questions after reading this, ask your healthcare provider. For more information, go to www.PanzygaInfo.com.

**What is PANZYGA?**

PANZYGA is a liquid medicine for infusion that contains immunoglobulin G (IgG), which are proteins that help fight infection. PANZYGA is used to treat primary humoral immunodeficiency (PI) in patients 2 years of age and older, chronic immune thrombocytopenia (cITP) in adults, and chronic inflammatory demyelinating polyneuropathy (CIDP) in adults.

PANZYGA is made from human plasma that is donated by healthy people and contains antibodies. For patients with PI, PANZYGA helps replace the missing antibodies in the body. For patients with cITP, PANZYGA helps the body produce more platelets (the blood cells that help blood clot) to control or prevent bleeding. For patients with CIDP, PANZYGA may help improve mobility and hand strength.

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- Nausea
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- Dermatitis
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- Abdominal pain
- Dizziness
- Anemia

These are not all the possible side effects. Talk to your HCP about any side effect that bothers you or that does not go away. Tell your HCP if you are pregnant, or plan to become pregnant, or if you are nursing. If you encounter any problems or experience side effects during or after the infusion, contact your healthcare provider. When doing so, keep your therapy tracker with you to be able to give all necessary information.

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This brief summary is based on the PANZYGA Prescribing Information (February 2021).

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# Ways to Give Back This Holiday Season

By Michelle Searle

**I CANNOT** believe the holidays are approaching once again! I'm sad to see summer go, and I'll be impatiently counting down the days until I can be back on the beach. But, while I hate the cold and not seeing the sun after 4 p.m., I do love the holidays! It's easy to get swept up in holiday shopping, movie marathons, stressing about finals or work that must be completed before a holiday break, traveling home and everything else the holidays bring. However, this is also the time to stop and think about others. And I don't just mean those for whom you're shopping. Having a primary immunodeficiency (PI), or any chronic illness, means we are part of a community. It might not have been a community we would have chosen for ourselves, but we weren't given a choice, so we must embrace it. The holidays are a great time to connect or reconnect with our chronic illness community; luckily for us, we can do it in several ways.

*Lend a helping hand to someone.* This year, when I attended the Immune Deficiency Foundation's (IDF) National Conference in Chicago, one of the speakers talked about having a buddy within the PI community who you can call if you need support, advice or accountability. It doesn't need to be someone who lives close to you, because just having someone who understands what you go through to text, call or FaceTime can make a world of difference. If you find a buddy who lives close, you can go on walks, grab a coffee or even do infusions together. Who says infusions need to be such a drag? If you'd like a buddy but need help finding one, join one of the Facebook groups for your condition, or join one of the online support groups IDF offers. If you're shy about asking a group of people (I know I would be), you could suggest the idea to the leaders of a support group you're a part of. Bring up the idea of buddies at the next meeting; I'm sure others in the group would love the idea and appreciate someone else bringing it up.

*Donate your time.* Foundations are always looking for volunteers and usually need help in many different ways. Some people offer their time as patient advocates. They help others learn more about PI, connect with the community and offer support. Another way you can help is by volunteering for a walk in your area or creating a walk if there isn't already one near you. Foundations also need volunteers for events throughout the year such as informational events, galas or support groups. For example, IDF offers many online support groups created and run by volunteers. There are groups in different cities, states, life stages and groups of people such as caregivers.

Another way you can volunteer your time is by visiting plasma centers. One of my previous columns discussed the rewarding experience of visiting plasma centers. I enjoy visiting these centers to talk to donors. When they meet someone who benefits from their plasma donations, it can impact how often they donate. There are many other ways to volunteer and donate your time. The first step is to visit the foundation's website where you want to volunteer. There, you can learn about the process of becoming a volunteer, time commitment obligations and the different ways you can volunteer. Helping others is very beneficial for our mental health. It can be challenging to find the time, but it's something I'm currently trying to do more often.

*Donate money.* The most basic and straightforward way to give back is to donate money. You can choose to donate to a foundation you care about, no matter how big or small the amount. For me, it would be IDF or the Make-A-Wish Foundation. I'm not going to be a hypocrite and suggest you ask people to donate to a foundation of your choice instead of giving you a holiday gift because, frankly, I look forward to my Christmas gifts! However, if you don't care about receiving gifts, asking people to donate is a great alternative. It can be as easy as setting up a donation page on your Facebook or Instagram page.

There are other ways to give back that I didn't include in this article. Even if you don't want to be involved in your chronic illness community, take a moment to reflect on how you can give back in other ways. If you're not a people person, there are a myriad of ways you can help that don't include people such as the environment or animals. We can all benefit from giving and receiving a helping hand. IG

**MICHELLE SEARLE** is a teacher from South Florida who was diagnosed with common variable immunodeficiency at 11 years old. She is currently living in New York where you will most likely find her eating pizza or trying to make friends with the local cats.

# Learning That "Good Enough" Is Good Enough

By Megan Ryan


**THIS YEAR** has been a tough one for me. My body decided it was not sufficiently "interesting enough" to my healthcare providers, and it wanted to show off and add some new medical complexities. As such, my team of physicians expanded with experts who slowly reached a consensus on my diagnosis and treatment plan. And that treatment plan required significant monitoring. My body was poked, scanned, prodded and scoped a record number of times, requiring me to bring all my resilience tools to the table. I also had to refresh my toolbox and my thinking box.

In the last few months, I've learned to accept that "good enough" *is* simply good enough in this season or in these parts of my life. I do not need to strive for perfection in everything I do. Until this point, I had always associated the words "good enough" with being inferior, subpar, mediocre or incompetent. And there's nothing about me, how I feel or what I do that is inferior, subpar, mediocre or incompetent. That's just not who I am.

So, it took some time for me to learn that "good enough" is about being satisfied, content or pleased. I have to silence my inner perfectionist and relish in the satisfaction of a priority task being accomplished. I now take pleasure in a simple meal prepared or the fact that leftovers from a meal will keep our family fed for a few days. I sent a birthday card with a handwritten note rather than attending an out-of-town party. I realize that most high school and college graduates and newly married couples just want cash as a gift, so there is no need for me to look for that elusive perfect gift. I'm focusing on being content with a task started and completed rather than a task done to a level of perfection that stays on my to-do list day after day after day.

In that same idea as "good enough," I have learned that "done is better than perfect." I realize that most people only read the first few sentences of an email message, so I changed my writing to be short and to the point at the top of the message and leave any details further below. I sent stacks of unread magazines to recycling or shared them with a neighbor rather than focusing on the fact that they were another "to do" on my list. I relocated the books on my to-be-read stack to a bookshelf rather than have them stare back at me every time I looked at the stack.

Reflecting on this, I realize that, for years, I put energy into tasks that did not require the energy that my body managing a complex chronic illness could give to a task. I exhausted myself trying to seek perfection when I needed to reserve and preserve energy and time for what really matters most: managing my health and meeting my basic needs. My focus on being "good enough" has brought much contentment to my days and that's now good enough for me!

**MEGAN RYAN** is a native Texan, lover of flowers, plants and gardening and always planning for an upcoming travel adventure. For more than 22 years, Megan has lived with common variable immune deficiency. She's taken her weekly treatments on the road to more than 20 countries and four continents so far.

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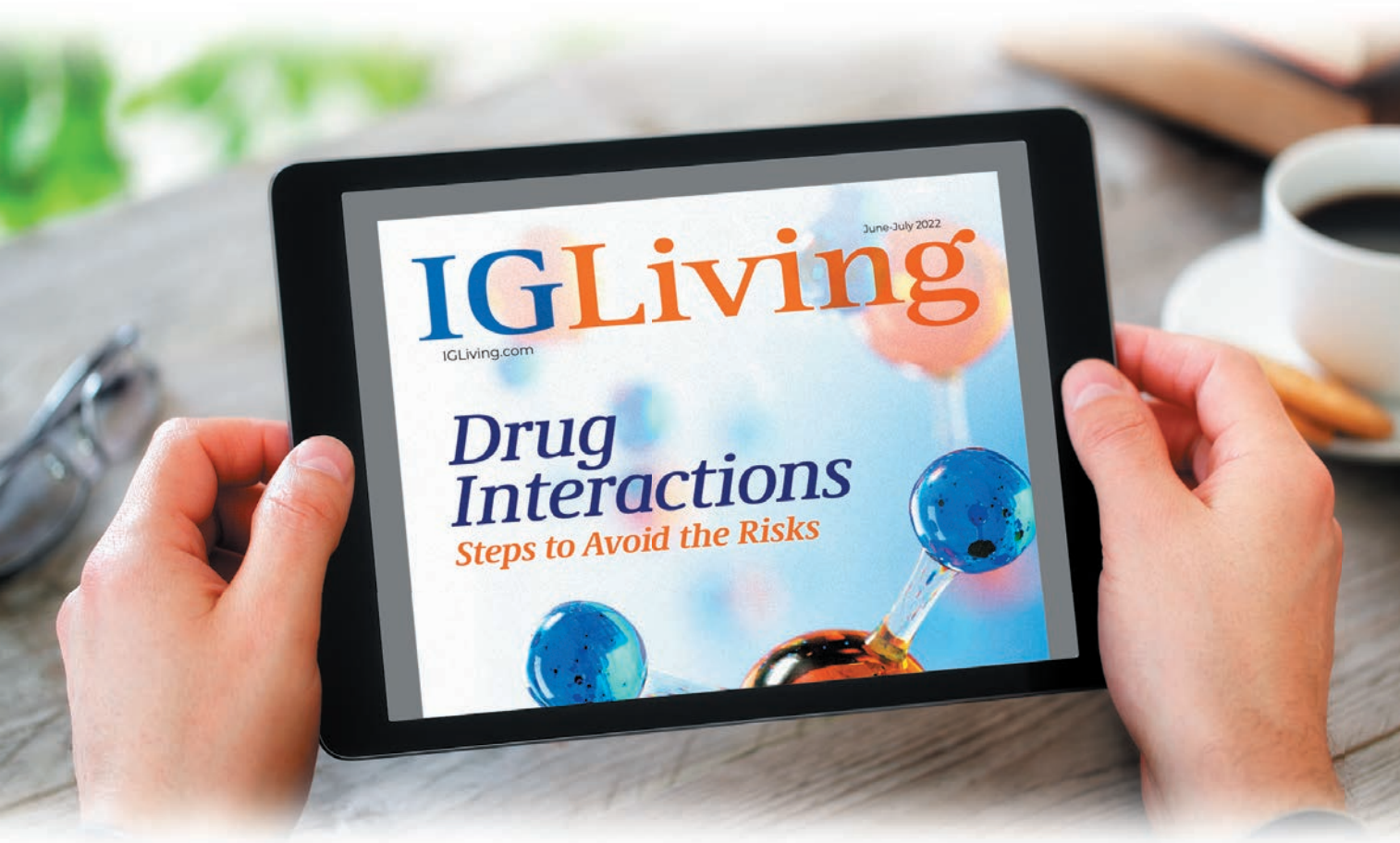


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# When Friendship Isn't Easy

By Jessica Leigh Johnson

**FRIENDSHIP IS** an integral part of childhood. Through friendships, children gain a sense of belonging and experience reduced loneliness and improved physical and mental well-being.[1] Unfortunately, children with chronic illness face unique challenges that can often make it hard to relate to and spend time with their peers, and sometimes finding and maintaining good friendships can be difficult.

## The Effects of Isolation

For children who suffer from primary immunodeficiency (PI), one of the main deterrents to friendship is isolation — not being able to take part in some of the social activities in which their peers are involved. The amount of social interaction or isolation children with PI experience greatly depends on the parents and their level of comfort with health-specific risk. Exposure to germs that pose a heightened risk of infection is one of the biggest threats to the health of children with PI, and less exposure to people and the outside world equals less exposure to germs. But, while this simplistic equation may result in greater physical health for children, it could be detrimental to their mental and social well-being.

It's been four years, but many of the negative effects of the COVID-19 pandemic still linger. Social health was one area of life that saw some of the most negative consequences of the lockdowns and isolating safety protocols. Many people reported increased feelings of loneliness during the pandemic,[2] and the reason is simple: Staying away from others to avoid sickness can make it more difficult to maintain friendships. What we learned on a grand scale from the pandemic is that isolation and separation are not good for us, socially or mentally. Unfortunately, these issues did not end after the pandemic for those who suffer from immune deficiencies. Life can often seem like a constant balancing act of risk vs. benefit for both children and their parents.

## When Pain or Immobility Limit Interactions

It's not always the fear of catching something and becoming ill that can limit children's in-person interactions with others. Sometimes it's just too painful or tiring to be anywhere but at home, and the idea of meeting friends for a bike ride or a game of volleyball can be daunting. Symptoms such as pain and fatigue, which can often be unpredictable, can make it challenging for young people living with chronic illnesses to take part in planned activities with their friends.[1] Because no one wants to cancel plans at the last minute when pain or fatigue strikes, many chronically ill teens may become hesitant to commit to plans with friends, knowing they may have to cancel. Sadly, their fear of disappointing their friends may lead them to withdraw, which in turn can cause friends to stop inviting them to activities altogether.[1]

## Some Advice for Friends of the Chronically Ill

Kids and teens with chronic illness may not want to burden their friends by asking them to make accommodations so they can be included. This definitely makes it more difficult for the

chronically ill to be an active part of their friend groups. But how can this be helped? One solution is to educate friends of the chronically ill about how to be encouragers, helpers, better communicators and all-around better friends.

Karissa Ewing, a teenager who suffers from a chronic condition called hereditary angioedema with normal C-1, or HAE, has created a list of practical ways to be a supportive friend to a child or teen with a chronic illness. Some of her suggestions are:[3]

• *Step inside their bubble.* Be present with your chronically ill friends, especially if they're isolating. Don't let them get lost in their bubble. Even if it's only for 15 minutes, do your best to visit when you have time.

• *Be persistent with communication.* "Whether it's a text, a phone call or a handwritten note, communication goes a long way in any friendship, especially a friendship with someone who has a chronic illness," Karissa says. "Note that your friend with a chronic illness may not be able to communicate as frequently or consistently when they are going through a tough time, but don't let that discourage you from reaching out and trying."

• *Be an encourager.* Kids and teens who suffer from chronic illness may deal with discouraging or even depressing thoughts. Their friends can serve as an encourager, reminding them of their strengths and listening when they share their feelings. Keep pointing them to what is true, and weed out any negative thoughts that may be the result of a bad day or hard time.

• *Check on them.* When people are first diagnosed with an illness, they receive more attention from friends and family. But after time goes by, others tend to move on with life, even if the child or teen is still suffering. Just because an illness is chronic, meaning it is always there, that doesn't mean it's any less difficult for the person dealing with it. So check on that friend frequently. Ask how he or she is and if you can do anything to help.

• *Don't lose empathy.* "No matter what, always remember that having a chronic illness is no easy feat," says Karissa. "Whenever your friend loses his or her temper, has a breakdown or simply doesn't want to talk, do your best to remind yourself it likely has nothing to do with you and everything to do with what [that friend is] going through. Try your best to put yourself in your friend's shoes, and don't lose empathy."

• *Remember you're only human.* Even if you're doing everything you can to ease the burden for a chronically ill friend, it's important to remember you can only do so much. As nice as it would be to have healing powers and magically remove the chronic illness from your friend's life, it's not possible. But, never underestimate the healing effects of simply being the best friend you can. Your chronically ill friend will surely notice and appreciate your efforts.

## Fostering Active and Healthy Relationships Is Possible

Children with chronic illness are no strangers to the limitations placed on them by their illness, but there is no need for their capacity for friendship to be limited as well. Kids who face adversity in life are quite resilient. They learn to find creative alternatives to problems that may seem overwhelming to others.[4] Thankfully, today's technology allows children and teens to stay connected to their friends without even being in the same location. With video calls, text messaging and social media, there are many ways to stay connected that may be less taxing for people with chronic illnesses.[1] With an Internet connection and a set of headphones, kids can play video games with their friends and communicate in real time from the comfort of their own homes. Obviously, this kind of interaction is inferior to in-person interaction (as we learned during the pandemic), but during times of illness or exhaustion, children and teens don't need to completely lose touch with their friend groups.

With the support of friends, family and healthcare professionals, children and teens with chronic illness can develop and foster active, healthy relationships with their friends. IG

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**JESSICA LEIGH JOHNSON** is a stay-at-home mom and mother of four kids, three of whom have X-linked agammaglobulinemia. She is a member of American Christian Fiction Writers and has written one book about the loss of her son to a primary immunodeficiency.

# A Stress-Free Spa Experience — at Home

By Rachel Maier, MS

**AH, SPA DAYS**! I love everything about them! The feel of plush, soft robes; the soothing scent of lavender and eucalyptus; the sight of low, flickering lights; the taste of the cucumber and mint-infused water — not to mention the spa services themselves. Facials, massages, manicures and pedicures? Sign me up for it all!

But regularly setting aside time and resources to indulge in professional spa days isn't realistic for me, and my guess is it might not be realistic for you either. Even splurging on at-home spa essentials can be a financial stretch!

## Stress — at the Spa?

While it's true that spas are designed to help us de-stress, the expense prohibits many of us from actually taking advantage of the treatments often enough to make a meaningful difference. Sometimes we do it anyway, telling ourselves we will figure out how to pay for it later, but the guilt for spending money we don't have and the anxiety about trying to scrape it together later hijacks our serenity and can leave us feeling worse than we did before!

Luckily, with a little planning and creativity, you can create your own soothing, stress-reducing spa experiences at home. Bonus? You can tailor them to your exact specifications and indulge in them as often as you like.

Be sure to make an appointment for your at-home spa day and mark it on your calendar. Protect that time. Treat it the same way you would treat a "real" spa appointment; otherwise, you might let life get in the way and not follow through.

## Invest in a Luxury Item

Set your budget, then set aside a portion of it to invest in a fancy spa must-have. Think about what you care about most and spend your money on it. For example, you might splurge on an indulgent, luxurious bath robe and pick up a bottle of Dr. Teal's lavender-scented foaming bath from Walmart (instead of breaking the bank on bougie bubbling bath from Oprah's most recent list of favorite things). Or, maybe you already have several skin care essentials but you would love to have a foot bath massager. A few other splurgy items are an essential oil diffuser and essential oils, a bubble massage bathtub mat, an electronic massage gun and a towel warmer.

## Purchase Affordable Items

Next, shop for items at a store where you can afford them. (Don't feel pressured to get spa essentials at an expensive boutique if you're on a dollar store budget.) If you're a DIY sort of person, your money may go farther by stocking up on ingredients such as unscented Epsom salt, baking soda, vegetable glycerin, coconut oil and two or three essential oils and making a variety of homemade skin care and bath products. A quick Internet search yields lots of free and easy skin care recipes.

## Use What You Have

You might be surprised by how many things you already have that will create a lovely spa experience at home. Use your best towels — fluff them in the dryer to freshen and warm them up. Fill up a pitcher with water, sliced cucumbers and mint; light your favorite candle; ask Siri to play spa music; fill up a clean dish pan with warm water and Epsom salts for a lovely foot soak; relax with sliced cucumbers on your eyes. What you already have is more than enough!

## Relax and Enjoy!

Make the time to find an at-home spa routine you love. It's worth it! It might take a little bit of planning and work up front, but once you find a routine you love, you'll be free to relax and enjoy. For more inspiration, check out the shopping guide. IG

**RACHEL MAIER,** MS, is the associate editor of *IG Living* magazine.


### *Dr. Teal's Relax & Relief Gift Set*

Rejuvenate your body and mind with this collection of spa-day essentials. Infused with eucalyptus and spearmint, these Dr. Teal's must-haves create a calming experience. Indulge your senses with an Epsom salt soak, foaming bath, body wash, bath and body oil, and body lotion. Dr. Teal's nourishing formulas are enriched with aloe vera, shea butter, vitamin E, sweet almond oil, jojoba oil and grapeseed oil so your skin can feel soft, moisturized and cared for.

*$25.49; www.amazon.com/Dr-Teals-Eucalyptus-Spearmint-Moisturizing/dp/B08Q2R8BL2*

### *Luxury Robe*

Splurge on a sumptuous Cozy Earth Luxe Bath Robe once and enjoy its spa-like luxury time and time again. Crafted from a luxurious blend of cotton and bamboo viscose using cutting-edge Zero Twist technology, this robe envelops you in unmatched softness and indulgence and is sure to be your new favorite part of spa day.

*$180; cozyearth.com/products/luxe-bath-robe?variant=43145103278260*

# Shopping Guide for an At-Home Spa Experience


### *Plant Therapy Pure Essential Oils*

Pure essential oils are a must-have for at-home spa experiences. Diffuse them to create a calming atmosphere in your home or add them to other basic skin care ingredients to create hand-crafted, custom spa products such as body scrubs, face masks and bath bombs. They help support a healthy immune system, revitalize your senses, promote relaxation and help relieve nervous tension. Not sure what set to start with? Try the lemon, lavender and peppermint bundle.

*Sets begin at $23.99; www.planttherapy.com/collections/sets*


### *Sweet Water Decor Spa Day Candle*

Captivating notes of sea salt, fresh jasmine, warm wood and cream will transport you to a relaxing place, leaving your stress behind. Crafted with clean-burning soy wax, this Spa Day candle creates a beautiful and spa-like environment wherever you are. Available in 9 ounce, 11 ounce and 15 ounce sizes.

*Starts at $19.99; www.sweetwaterdecor.com/collections/spa-day*

### *Facial Steamer*

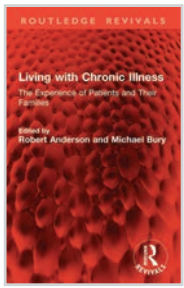
Up your at-home facial game with the Plum Beauty Ionic Facial Steamer. Your skin will take on a natural, healthy glow in a matter of minutes. The steamer opens up pores allowing for deep cleansing; hydrates and detoxifies skin; promotes a radiant and refreshed complexion; and allows for better absorption of skin care products.

*$34.99; myplumbeauty.com/product/ionic-facial-steamer*

### *Lotion Warmer*

Give your skin an indulgent treat with this heated lotion dispenser. Fill the reservoir with your favorite hand or body lotion, wait two minutes for it heat up and then massage warm lotion into your skin for a soothing spa experience.

*$29.99; www.conair.com/en/heated-lotion-dispenser/HLD23TGR.html*

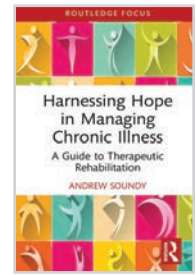



### *Living with Chronic Illness: The Experience of Patients and Their Families (Routledge Revivals), 1st Edition*

*Editors: Robert Anderson and Michael Bury*

*Publisher: Routledge*

First published in 1988, *Living with Chronic Illness* presents a vivid account of the reality of life with chronic illness from the perspective of patients and their families. The authors look at the expectations, priorities and problems of those most affected by chronic illness, and examine the strategies they have developed to cope with their considerable disadvantages. The experience of carers and the ways in which their problems change over time are major themes in the book. An overview of the consequences of particular illnesses are presented, before discussion of specific problems in daily life — maintaining family relationships, managing treatment regimens, coping with work and home commitments, and living with bodily change and social stigma.



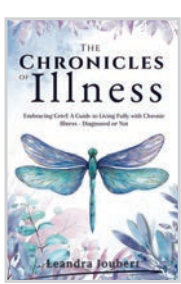

### *Harnessing Hope in Managing Chronic Illness: A Guide to Therapeutic Rehabilitation, 1st Edition*

*Author: Andrew Soundy*

*Publisher: Routledge*

This book provides a new framework to enable physiotherapists and other healthcare professionals to engage with patients to create better interactions and outcomes for rehabilitation. Based on extensive research into how patients express their experiences, it identifies those factors that influence how hope can be used to benefit an interaction. It also considers central questions to illustrate how interactions can be psychologically mapped to assess emotions, adjustment and hope. The book then features practical guidance on how to integrate the idea of hope into therapeutic conversations with patients, fostering acceptance and adaptation to the present, and looking toward the future.

# New and Useful Reading




### *The Chronicles of Illness: Embracing Grief: A Guide to Living Fully with Chronic Illness, Diagnosed or Not*

*Author: Leandra Joubert*

*Publisher: Independently Published*

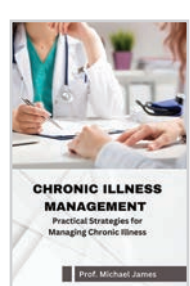
*The Chronicles of Illness* is a practical guide for anyone navigating the complex emotions and challenges of living with a chronic illness, whether diagnosed or not. Written by a specialist wellness counsellor who shares personal experiences and insights, this book offers tools to help readers find strength and self-worth amid their health challenges. Provided are practical strategies and heartfelt advice to manage chronic illness's emotional and physical aspects. At its core is the belief that a diagnosis does not define a person. It may change certain aspects of life, but it does not diminish one's value or capacity to enjoy life.

### *Chronic Illness Management: Practical Strategies for Managing Chronic Illness*

*Author: Professor Michael James*

*Publisher: Independently Published*

This book contains practical advice and proven strategies to help patients manage their condition effectively. Included is information about understanding diagnoses to building supportive healthcare teams, developing personalized management plans and integrating nutrition, exercise and mental health practices. Also included are tips for balancing work, family and social life, navigating daily challenges and exploring alternative and complementary therapies.



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### Ataxia Telangiectasia (A-T)

**Websites**

- A-T Children's Project: www.atcp.org

### Chronic Inflammatory Demyelinating-Polyneuropathy (CIDP)

**Websites**

- GBS/CIDP Foundation International: www.gbs-cidp.org

### Evans Syndrome

**Online Peer Support**

- Rare Connect Evans Syndrome Community Group: www.rareconnect.org/en/community/evans-syndrome/faqs

### Guillain-Barré Syndrome (GBS)

**Websites**

- GBS/CIDP Foundation International: www.gbs-cidp.org
- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

**Online Peer Support**

- GBS Support Group: www.gaincharity.org.uk
- GBS/CIDP Foundation International Community Forums: forum.gbs-cidp.org

### Immune Thrombocytopenia (ITP)

**Websites**

- ITP Support Association – UK: www.itpsupport.org.uk
- Platelet Disorder Support Association: www.pdsa.org

### Kawasaki Disease

**Websites**

- American Heart Association: www.heart.org/en/health-topics/kawasaki-disease
- American Academy of Family Physicians: www.aafp.org/afp/2006/1001/p1141.html
- Kawasaki Disease Foundation: www.kdfoundation.org
- KidsHealth: www.kidshealth.org/parent/medical/heart/kawasaki.html

### Mitochondrial Disease

**Websites**

- United Mitochondrial Disease Foundation: www.umdf.org
- MitoAction: www.mitoaction.org

### Multifocal Motor Neuropathy (MMN)

**Websites**

- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

### Multiple Sclerosis (MS)

**Websites**

- Multiple Sclerosis Association of America: www.mymsaa.org
- Multiple Sclerosis Foundation: www.msfocus.org
- National Multiple Sclerosis Society: www.nationalmssociety.org

**Online Peer Support**

- Friends with MS: www.FriendsWithMS.com
- MSWorld's Chat and Message Board: www.msworld.org
- Overcoming Multiple Sclerosis: www.overcomingms.org/community

### Myasthenia Gravis (MG)

**Websites and Chat Rooms**

- Myasthenia Gravis Foundation of America (MGFA): www.myasthenia.org
- Myasthenia Gravis Association: mgakc.org

**Online Peer Support**

- Genetic Alliance: www.geneticalliance.org

### Myositis

**Websites**

- The Myositis Association: www.myositis.org
- International Myositis Assessment and Clinical Studies Group: www.niehs.nih.gov/research/resources/imacs/index.cfm

**Online Peer Support**

- Juvenile Myositis Family Support Network: www.curejm.org/fsn/index.php
- The Cure JM Foundation: www.curejm.org
- Myositis Association Support Group: www.myositis.org/patient-support/support-groups
- Myositis Support Group – UK: www.myositis.org.uk

### Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS)

**Websites**

- PANS/PANDAS UK: www.panspandasuk.org
- PANDAS Network: www.pandasnetwork.org
- PANDAS Physician Network Family Resources: www.pandasppn.org/parent-information
- National Institute of Mental Health: www.nimh.nih.gov/health/publications/pandas/index.shtml

### Pemphigus and Pemphigoid

**Websites**

- The International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

### Peripheral Neuropathy (PN)

**Websites**

- Neuropathy Action Foundation: www.neuropathyaction.org
- Western Neuropathy Association: www.pnhelp.org
- Neuropathy Alliance of Texas: www.neuropathyalliancetx.org
- The Foundation for Peripheral Neuropathy: www.foundationforpn.com

### Primary Immune Deficiency Disease (PI)

**Websites**

- Immune Deficiency Foundation: www.primaryimmune.org
- Jeffrey Modell Foundation: www.info4pi.org
- The National Institute of Child Health and Human Development (NICHD): www.nichd.nih.gov/Pages/index.aspx
- American Academy of Allergy, Asthma & Immunology: www.aaaai.org
- International Patient Organisation for Primary Immunodeficiencies (IPOPI) — UK: www.ipopi.org
- Rainbow Allergy-Immunology: www.uhhospitals.org/rainbow/services/pediatric-allergy-and-immunology

**Online Peer Support**

- IDF Friends: www.idffriends.com
- Jeffrey Modell Foundation Facebook Page: www.facebook.com/JMFworld
- IDF Peer Support Program: www.primaryimmune.org/idf-peer-support-program

### Scleroderma

**Websites**

- Scleroderma Foundation: www.scleroderma.org
- Scleroderma Research Foundation: www.srfcure.org
- Johns Hopkins Scleroderma Center: www.hopkinsscleroderma.org

**Online Peer Support**

- Scleroderma Support Forum: www.curezone.com/forums/f.asp?=404

### Stiff Person Syndrome (SPS)

**Websites**

- American Autoimmune Related Diseases Association Inc.: www.aarda.org
- Genetic Alliance: www.geneticalliance.org
- Living with Stiff Person Syndrome (personal account): www.livingwithsps.com
- The Stiff Person Syndrome Research Foundation: stiffperson.org

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